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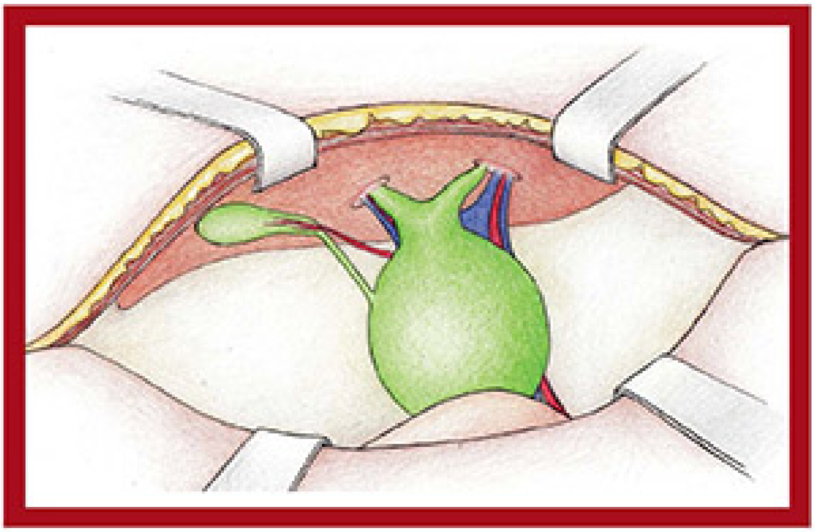

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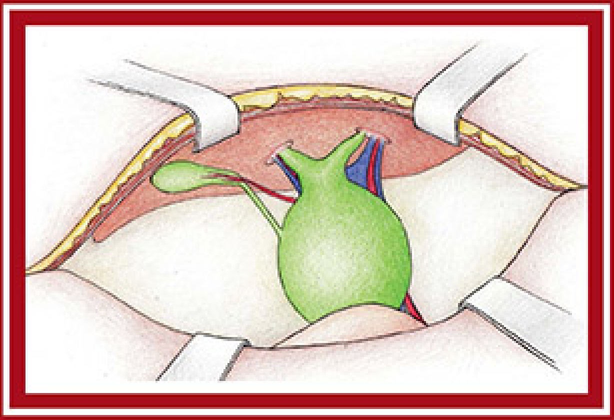

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Preface

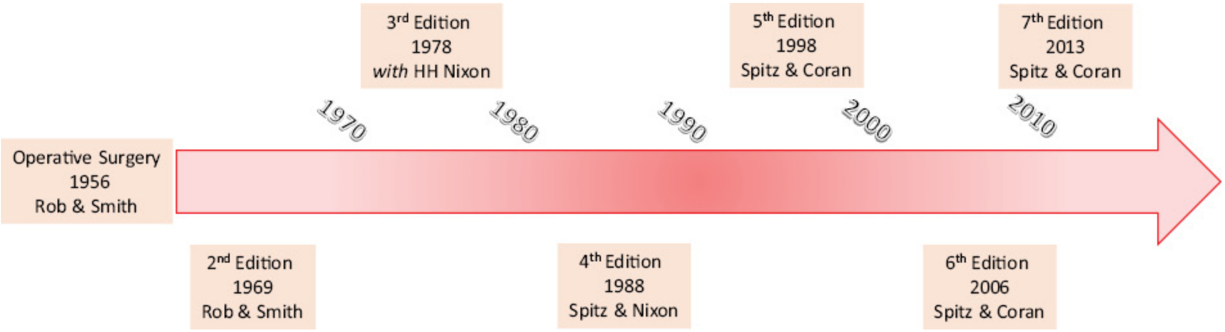
Charles Granville Rob (1913–2001), an English vascular surgeon who became Professor of Surgery in Rochester, New York, and Rodney (later Lord) Smith (of Marlow) (1914–1998), a London hepatobiliary surgeon, published and oversaw the first three editions of *Operative Surgery* from 1956. Over the next 20 years or so they proceeded to co-write and edit a suite of surgical specialty textbooks devoted to technique and operations. Mr. Harold Homewood Nixon from Great Ormond Street in London became the first specialist pediatric collaborator and, in the fourth edition, he took on the editorial role in combination with Lewis Spitz from the same hospital. Lewis was joined by Arnold Coran from Michigan from the fifth edition onward, with a noticeable lurch across the Atlantic in terms of the increased proportion of invited North American authors and use of American spelling (Figure 1).

The new editors for this eighth edition are Mark Davenport, from London, and James Geiger, from Michigan, both of whom have had privileged mentor relationships with their predecessors. This edition offers a break with the past and some serious revision of contents and presentation. We have tried to simplify and, to some extent, condense the previous structure with its 100+ chapters by combining open and endoscopic / minimally invasive surgical procedures in single chapters, often with two authors from either side of the great divide. Our reasoning has been that the condition / disease is the same although the route to successful therapy may well be different. We have tried to retain key surgeons who are recognized throughout the world as masters of their art – and this is certainly obvious in operations requiring the highest degrees of skill or judgment.

The most obvious change, though, is in terms of presentation. This edition makes significant use of color in the schematics and anatomical artwork, sees the introduction of operative photographs and, most appositely, includes a bank of reference videos, which we hope will aid understanding while demonstrating real life in the operating room.

As with all great writing endeavors, there has been a great deal going on in the background, unseen and probably uncredited. This is, after all, the culmination of the work of a host of international surgeons with different styles and accomplishments but all united in the desire to produce the best product they can. We hope that the final version is more than the sum of its parts. To achieve this, we required considerable help from our associate editors, Nigel Hall and Steven Rothenberg, the latter hosting a memorable editorial meeting at the onset of winter in his Eagles' Nest in Colorado. We would also like to thank Samantha Cook and Miranda Bromage in the office of the publishers for their considerable professional insights and input.

James Geiger dedicates this work to Daniel H. Teitelbaum, MD (1956–2016). Dan served as an Associate Editor of the seventh edition of *Operative Pediatric Surgery*. Had he not sadly passed away at the age of 59 years, after a long struggle with a brain tumor, he would have been one of the editors of this edition. Dan was a brilliant clinician and scientist who made signification contributions to improving the care of children with short bowel syndrome, Hirschsprung disease, and inflammatory bowel disease. He was loved greatly by his patients, who recognized his great skill and selfless devotion to their care, and also by the residents, fellows, and colleagues, who valued his great wisdom, intellect, and kindness. Dan's extraordinary accomplishments will forever impact the care of children.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1 Timeline of Operative Pediatric Surgery.

Mark Davenport dedicates this work to Derek (1927–2018) and Gladys (1927–2019) Davenport.

Mark Davenport, ChM, FRCS(Paeds), FRCS(Eng)
James D. Geiger, MD, FACS

List of abbreviations used

ACC	adrenocortical carcinoma
ACE	antegrade colon enema; antegrade continence enema
ACT	activated clotting time
AFP	alpha-fetoprotein
AH	axillary hyperhidrosis
ALF	acute liver failure
ALT	alanine transaminase
AMH	anti-Müllerian hormone
ANC	absolute neutrophil count
APSA	American Pediatric Surgical Association
ARM	anorectal malformation
ARMS	alveolar rhabdomyosarcoma
ASIS	anterior superior iliac spine
AST	aspartate transaminase
ATA	American Thyroid Association
ATLS	Advanced Trauma Life Support®
AUA	American Urological Association
AVF	arteriovenous fistula
AVM	arteriovenous malformation
β-HCG	beta-human chorionic gonadotropin
BA	biliary atresia
BAPS CASS	British Association of Paediatric Surgeons Congenital Anomaly Surveillance System

BAPU	British Association of Paediatric Urologists
BMI	body mass index
BRBNS	blue rubber bleb nevus syndrome
BWS	Beckwith–Wiedemann syndrome
BXO	balanitis xerotica obliterans
CAH	congenital adrenal hyperplasia
CAM	cystic adenomatoid malformation
CBC	complete blood count
CBD	common bile duct
CCAM	congenital cystic adenomatoid malformation
CCM	congenital choledochal malformation
CD	Crohn's disease
CDAD	*Clostridium difficile*-associated disease
CDH	congenital diaphragmatic hernia
CDHSG	Congenital Diaphragmatic Hernia Study Group
CDI	*Clostridium difficile* infection
CF	cystic fibrosis
CFTR	cystic fibrosis transmembrane conductance regulator
CGRP	calcitonin gene-related peptide
CHAOS	congenital high airway obstruction syndrome
CHD	common hepatic duct
CLOVES	congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and skeletal anomalies
CM	capillary malformation
CMV	cytomegalovirus
CNB	core needle biopsy
CNS	central nervous system
COG	Children's Oncology Group
COPUM	congenital obstructive posterior urethral membrane
CPAM	congenital pulmonary airway malformation
CPAP	continuous positive airway pressure
CRBSI	catheter-related bloodstream infection
CRE	controlled radial esophageal

CRP	C-reactive protein
CRS	cytroreductive surgery
CS	compensatory sweating; compensatory swelling
CSF	cerebrospinal fluid
CT	computed tomography
CUSA	cavitron ultrasonic surgical aspirator
CVC	central venous catheter
CVP	central venous pressure
CXR	chest X-ray
DCS	dynamic compressor system
DEXA	dual energy X-ray absorptiometry analysis
DJ	duodenojejunal
DMSA	dimercaptosuccinic acid
DPE	delayed primary excision
DPL	diagnostic peritoneal lavage
DSD	disorders of sexual development
DSR	distal splenorenal
DSRS	distal splenorenal shunt
DVT	deep vein thrombosis
DWR	diameter/weight ratio
Dx/HA	dextranomer/hyaluronic acid
EA	esophageal atresia
EAU	European Association of Urology
EBV	Epstein–Barr virus
ECG	electrocardiogram
ECLS	extracorporeal life support
ECMO	extracorporeal membrane oxygenation
EEA	end-to-end anastomosis
EFS	event-free survival
EGD	esophagogastroduodenoscopy
EGJ	esophagogastric junction
EHL	electrohydraulic lithotripsy
EHPVO	extrahepatic portal vein occlusion

ELBW	extremely low birth weight
EMR	endomucosal resection
ENT	ear, nose, and throat
ERAS	enhanced recovery after surgery
ERCP	endoscopic retrograde cholangiopancreatography
ERDS	endorectal double-stapled
ERMS	embryonal rhabdomyosarcoma
ESPGHAN	European Society for Pediatric Gastroenterology, Hepatology, and Nutrition
ESWL	extracorporeal shock-wave lithotripsy
ETT	endotracheal tube
EXIT	*ex utero* intrapartum treatment
FAST	focused abdominal sonography for trauma
FDA	Food and Drug Administration
FEV1	forced expiratory volume in the first second
FFS	failure-free survival
FISH	fluorescent *in situ* hybridization
FNH	focal nodular hyperplasia
FVC	forced vital capacity
GA	general anesthesia
GALT	gut-associated lymphoid tissue
GCST	granulocyte colony stimulating factor
GE	gastroesophageal
GERD	gastroesophageal reflux disease
GI	gastrointestinal
GIR	intravenous glucose infusion rate #
GJ	gastrojejunostomy
GU	genitourinary
GVHD	graft versus host disease
HB	hepatoblastoma
HCC	hepatocellular carcinoma
hCG	human chorionic gonadotropin
HCN-NOS	hepatocellular neoplasm-not otherwise specified
HI	hyperinsulinism

HIDA	hepatobiliary iminodiacetic acid
HIPEC	hyperthermic intraperitoneal chemotherapy
HIT	hydrodistension implantation technique
HIV	human immunodeficiency virus
HME	heat and moisture exchange
HPVG	hepatic venous pressure gradient
HU	Houndsfield unit
IBD	inflammatory bowel disease
IBD-U	inflammatory bowel disease-unspecified
IC	ileocecal
ICD	intercostal drain
ICG	indocyanine green
ICU	intensive care unit
IDA	iminodiacetic acid
IDRF	image-defined risk factor
IFALD	intestinal failure-associated liver disease
IHH	infantile hepatic hemangioma
IJV	internal jugular vein
IMRT	intensity-modulated radiation therapy
IMV	inferior mesenteric vein
iNO	inhaled nitric oxide
INoEA	International Network of Esophageal Atresia
INR	international normalized ratio
INRGSS	International Neuroblastoma Risk Group Staging System
INSL3	insulin-like hormone 3
INSS	International Neuroblastoma Staging System
IPAA	ileal pouch–anal anastomosis
IR	interventional radiology
IRA	ileorectal anastomosis
ISS	injury severity score
ISSVA	International Society for the Study of Vascular Anomalies
IVC	inferior vena cava
IVU	intravenous urography

KPE	Kasai portoenterostomy
LAARP	laparoscopic assisted anorectal pull-through
LESS	laparoendoscopic single-site surgery
LHR	lung to head ratio
LILT	longitudinal intestinal lengthening and tailoring
LINE	localized islet nuclear enlargement
LM	lymphatic malformation
LMA	laryngeal mask airway
LMIC	low- and middle-income countries
LOF	loss of function
LT	liver transplantation
LUQ	left upper quadrant
MAGPI	meatal advancement and glanuloplasty
MCDK	multicystic dysplastic kidney
MCT	medium-chain triglyceride
MELD	model for endstage liver disease
MEN	multiple endocrine neoplasia
MG	myasthenia gravis
MGD	mixed gonadal dysgenesis
MIBG	metaiodobenzylguanidine
MID	minimal important difference
MII	multiple intraluminal impedance
MIP	megameatus-intact prepuce
MIS	Müllerian inhibiting substance
MKI	mitosis–karyorrhexis index
MMC	myelomeningocele
MOMS	Management of Myelomeningocele Study
MRA	magnetic resonance angiography
MRB	meso-Rex bypass
MRCP	magnetic resonance cholangiopancreatography
MRE	magnetic resonance elastography
MRI	magnetic resonance imaging
MRU	magnetic resonance urography

MRV	magnetic resonance venography
MSC	mesocaval shunt
NASPGHAN	North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition
NEC	necrotizing enterocolitis
NI	neurological impairment
NIRS	near-infrared spectroscopy
NSAID	non-steroidal anti-inflammatory drug
NSS	nephron-sparing surgery
NWTSG	North American National Wilms' Tumor Study Group
o/e	observed-to-expected
PAS	preauricular sinus
PC	pressure of correction
PCCL	percutaneous cystolithotomy
PCDAI	Pediatric Crohn's Disease Activity Index
PCNL	percutaneous nephrolithotomy
PCR	polymerase chain reaction
PDA	patent ductus arteriosus
PDS	polydioxanone suture
PE	pulmonary embolism
PEEP	positive end-expiratory pressure
PEG	percutaneous endoscopic gastrostomy
PELD	pediatric endstage liver disease
PEM	peroral endoscopic myotomy
PEP	post-ERCP pancreatitis
PET	positron emission tomography
PFIC	progressive familial intrahepatic cholestasis
PH	primary hyperhidrosis
PHT	portal hypertension
PICC	peripherally inserted central venous catheter
PMD	pressure measuring device
PN	parenteral nutrition
POEM	peroral endoscopic myotomy
POST-TEXT	post-treatment extent of disease

PPD	primary peritoneal drainage
PPE	parapneumonic effusion
PPH	primary palmar hyperhidrosis
PPI	proton pump inhibitor
PPV	patent processus vaginalis
PR	per rectum
PRE	primary re-excision
PRETEXT	pre-treatment extent of disease
PS	percutaneous stay-sutures
PSARP	posterior sagittal anorectoplasty
PSARVUP	posterior sagittal anorectovaginourethroplasty
PSI	pounds per square inch
PSS	portosystemic shunt
PT	pressure of treatment; prothrombin time
PTC	percutaneous transhepatic cholangiography
PTFE	polytetrafluoroethylene
PTH	parathyroid hormone
PTT	partial thromboplastin time
PUCAI	Pediatric Ulcerative Colitis Activity Index
PUJ	pelviureteric junction
PUM	partial urogenital mobilization
PUV	posterior urethral valve
PV	processus vaginalis
PVC	polyvinyl chloride
RAL	robotic-assisted laparoscopy
RC	right carotid
RI	reflux index
RICH	rapidly involuting congenital hemangioma
RIJV	right internal jugular vein
RIRS	retrograde intrarenal surgery
RLN	recurrent laryngeal nerve
RMS	rhabdomyosarcoma
RP	restorative proctocolectomy; retroperitoneoscopic

RT	radiotherapy
RUQ	right upper quadrant
RUTI	recurrent urinary tract infection
RYGBP	Roux-en-Y gastric bypass
SBO	short bowel obstruction
SBS	short bowel syndrome
SCD	sickle cell disease
SCID	severe combined immunodeficiency
SCM	sternocleidomastoid muscle
SCT	sacrococcygeal teratoma
SG	sleeve gastrectomy
SIAA	straight ileoanal anastomosis
SIDS	sudden infant death syndrome
SIMPL	single-instrument port laparoscopic nephrectomy
SIOP	Société Internationale d'Oncologie Pédiatrique
SIP	spontaneous intestinal perforation
SLO	second-look operation
SLPC	selective laser photocoagulation
SMA	superior mesenteric artery
SMP	super mini PCNL
SMV	superior mesenteric vein
SSEP	somatosensory evoked potential
SSI	surgical site infection
STEP	serial transverse enteroplasty (procedure)
STL	stereolithography
STS	sodium tetradecyl sulfate
SVC	superior vena cava
TBSA	total body surface area
TDD	thoracoscopic debridement and decortication
TEF	tracheoesophageal fistula
TEGD	total esophagogastric dissociation
TFLV	total fetal lung volume
TI	terminal ileum

TIP	tubularized incised plate
TIPS	transjugular intrahepatic portosystemic shunt
TNF	tumor necrosis factor
tPA	tissue plasminogen activator
TP	transperitoneal
TPN	total parenteral nutrition
TS	thoracic sympathectomy; trauma score
TSH	thyroid stimulating hormone
TTTS	twin-to-twin transfusion syndrome
TUM	total urogenital sinus mobilization
TUR	transurethral resection
UC	ulcerative colitis
UESL	undifferentiated embryonal sarcoma of the liver
UGI	upper gastrointestinal
UGS	urogenital sinus
UNOS	United Network for Organ Sharing
UPJ	ureteropelvic junction
URS	ureterorenoscopy
US	ultrasonography; ultrasound
UTI	urinary tract infection
UW	University of Wisconsin
VA	venoarterial; vincristine and actinomycin-D
VAC	vincristine, actinomycin-D, and cyclophosphamide
VATS	video-assisted thoracic surgery; video-assisted thoracoscopy
VCUG	voiding cystourethrogram
VIP	vasoactive intestinal polypeptide
VM	venous malformation
VSD	ventricular septal defect
VUJ	vesicoureteric junction
VUR	vesicoureteric reflux
VURD	unilateral renal dysplasia
VV	venovenous
WBC	white blood cell

WHO	World Health Organization
WT	Wilms' tumor
YAG	yttrium aluminium garnet

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Vascular access

MARCUS D. JARBOE AND RONALD B. HIRSCHL

HISTORY

Secure vascular access is central to the delivery of many aspects of modern medical care. In 1628, William Harvey described the heart and circulation in detail. This provided the anatomical basis for subsequent vascular interventions. By the early 1900s, intravenous (IV) therapy was becoming established, replacing proctoclysis and cutaneoclysis as a means of delivering fluids and drugs to patients. Effective antimicrobials, chemotherapeutic agents, parenteral nutrition, and the evolution of a culture of intensive care are among the medical advances that have encouraged the development of innovative strategies for vascular access in children over the last 60 years. This group of procedures is now one of the most common performed by pediatric surgeons.

PRINCIPLES AND JUSTIFICATION

Venous and arterial access is a key part of the management of many children who require investigations, monitoring, and specific interventions. Access procedures may facilitate the monitoring of physiological, hematological, and biochemical indices. Therapeutic indications include the delivery of fluids, blood products, nutrition, and drugs, hemodialysis as well as miscellaneous interventions such as endovascular surgery and, increasingly, cellular transplantation.

Some children put forward for vascular access may be suitably managed by simpler, less invasive, and equally effective means. Despite improvements in devices, insertion techniques, and postoperative care, distress and complications related to catheter insertion are not infrequent and may be severe. In consultation with other members of the multidisciplinary team, it is the responsibility of the surgeon to ensure that the vascular access procedure proposed for each child can be justified.

OVERVIEW OF DEVICES AND TECHNIQUES

In most children, short-term venous access for sampling and the delivery of non-irritant infusions is achieved by the use of a short 14- to 26-gauge cannula inserted into a superficial vein in an upper or lower limb. In addition to easily visible veins, useful sites include the long saphenous vein anterior to the medial malleolus at the ankle, the cephalic vein at the wrist, the interdigital vein between the fourth and the fifth metacarpals on the dorsum of the hand, the external jugular vein, the superficial temporal vein of the scalp in small infants, and, occasionally, superficial veins on the trunk. Generally, the main role of the pediatric surgeon is related to central venous access. However, if the surgeon has reasonably developed ultrasound skills, they can play a large role in peripheral access in the difficult patient population in the operating room environment.

The large number of central venous access devices fall into two broad categories:

- *Non-tunneled catheters*. These include percutaneously inserted central venous catheters (PICCs), percutaneous central venous catheters (CVCs), and acute hemodialysis catheters.
- *Tunneled catheters*. Examples included cuffed CVCs (such as Hickman, broviacs or hemodialysis catheters) and venous port devices.

The type of central venous access device used will depend on the requirements of the individual child (**Table 1.1**).

Table 1.1 **Types of central venous access device**

Device	Characteristics	Advantages	Disadvantages	Typical indications
Non-tunneled CVC	Short, relatively stiff, usually multiple-lumen catheter	Ease of insertion	High infection rate	Short-term IV therapy, pressure monitoring, "bridge access" before longer lasting line
Single-lumen Hickman catheter	Relatively soft catheter	Relatively low infection rate	No possibility of coadministration of incompatible infusions	Total parenteral nutrition, low intensity chemotherapy (e.g. nephroblastoma)
Multiple-lumen Hickman catheter	Relatively soft catheter with two or three lumens	Coadministration of blood products, parenteral nutrition, and drugs	Higher infection rate	Intensive chemotherapy protocols, bone marrow transplantation
Non-cuffed tunneled CVC	Small-caliber soft catheter with one or two lumens	Ease of removal	Higher rate of inadvertent removal and infection	Short- to medium-term access with reliable blood sampling
Non-tunneled hemodialysis catheter	Large diameter catheter with offset lumens	Ease of insertion	Short lifespan, higher infection rate	Short-term hemodialysis, plasmapheresis, stem cell harvest
Tunneled (permanent) hemodialysis catheter	Large diameter catheter with offset lumens	Long lifespan, low infection rate	Higher incidence of damage to vein	Long-term hemodialysis
Central venous port device	Subcutaneous port with attached venous catheter	Even lower infection rate than Hickman catheter	Requires needle access, longer scar than Hickman catheter, visible "port bump"	Chemotherapy, conditions requiring regular transfusions of blood products (e.g. hemophilia) or antibiotics (e.g. cystic fibrosis)
PICC	Small-caliber soft catheter with one or two lumens	Safe insertion without general anesthetic, ease of removal	Higher rate of inadvertent removal and occlusion	Short- to medium-term access (e.g. for antibiotic therapy)

Emergency intraosseous access and arterial cannulation for monitoring are described in the section "Emergency intraosseous venous access," but other vascular procedures such as endovascular surgery, and extracorporeal membrane oxygenation are beyond the scope of this chapter.

PREOPERATIVE

Assessment and planning

A specific history and clinical examination are imperative, especially if there has been previous central venous cannulation. This should include the proposed site of insertion and exit site of the device and a search for stigmata that suggest potential venous access difficulty, such as multiple scars and dilated body wall veins. If such stigmata exist, preoperative vascular imaging (e.g. magnetic resonance venography (MRV), venogram, and ultrasound) may be indicated.

Consent should be obtained by the operating surgeon or a colleague with a clear understanding of the procedures and devices involved, as well as possible complications. Coagulation parameters should be optimized prior to surgery. With proper planning, most of these procedures can be carried out electively, reducing risk to patients by a reduction of afterhours operating.

Central venous access sites

For the purposes of central venous access, the central veins may be defined as the superior vena cava (SVC), the right atrium, and the suprahepatic inferior vena cava (IVC). In general, the right internal jugular vein (RIJV) is the best site for central venous access. Other potential veins, in the usual order of preference, are the left internal jugular, external jugular, axillary, common femoral, and subclavian veins. With ultrasound-guided puncture, this may expand to other veins, such as the brachiocephalic veins or even the SVC from the supraclavicular approach. Less conventional methods, such as recanalization of occluded veins, the use of small collateral veins, and transhepatic or translumbar access to the IVC are occasionally required. These should be undertaken by personnel skilled in image-guided techniques and are outside the scope of this text.

Central catheter tip location

The ideal position of the upper CVC tip is contentious. The options include the SVC, the right atrium, and at the junction between the two. Individual patient requirements, government guidelines, manufacturer's recommendations along with institutional and personal experience and practice should be taken into consideration when deciding the final position. In general, our preference is for placement in the SVC–right atrial junction or proximal right atrium. The catheter tip should not be deep enough to injure the valve or atrial wall and should be distal to the brachiocephalic veins. In the IVC, true central location is after the confluence of the hepatic vein and the IVC (suprahepatic IVC). Lines shorter can result in extravasation or vessel injury from noxious medications/infusions. However, lines this long may not be optimal for rapid infusion resuscitation.

ANESTHESIA

Anesthetic should be appropriate to the complexity of the child undergoing surgery. Postoperative management may require intensive care facilities, which should be arranged in advance.

Anesthetic strategies for vascular access procedures in children differ widely between centers. IV sedation may be used with good results. Our institutional preference is for general anesthesia, but selected children will tolerate CVC insertion under local anesthetic with or without an inhalational agent.

In the majority of cases, the airway is best secured by muscle paralysis and an endotracheal tube. A laryngeal mask may be appropriate for some patients if muscle relaxants are not used. Slight head-down positioning and continuous positive airway pressure are advisable to reduce the risk of air embolus. Standard monitoring includes oxygen saturation, end-tidal carbon dioxide, temperature, blood pressure, and electrocardiography. The airway must be monitored closely, especially each time the position of the head is altered. Injection of local anesthetic at the site of all incisions is recommended. This should be done before incisions are made as it allows for a lighter general anesthetic and eliminates the risk of damaging the device following implantation.

EQUIPMENT

Access

The needle used for vascular access varies according to size of the vessel. Most vessels can be accessed with 21-gauge or 22-gauge micropuncture needles. Small vessels such as basilic veins or radial artery in a premature neonate may require needles as small as 26 gauge. Hollow-bore single puncture needles that are 21 gauge and smaller are sometimes called micropuncture needles and are exceptionally safe to use. Inadvertent puncture of surrounding structure rarely results in any problems given the small size. Larger needles, such as 18 gauge, carry significant risk of damage to nearby structure and the target vein itself. A 22-gauge needle allows passage of a 0.018-inch (0.5 mm) coaxial wire. In the authors' practice, the 0.018-inch Cope nitinol Mandril Wire Guide (Cook Medical, Bloomington, IN) is preferred. This wire has a flexible tip, which reduces risk of vascular damage from wire passage, but also a stiff shaft, which increases ease of dilator and catheter threading. Smaller needles and angiocaths require smaller diameter wires such as 0.010 inch.

Wires are often useful in vascular access but in small children and infants J-wires or C-wires should be avoided. The tips of these wires form a safe, blunt curl at the front end of the wire as they leave the needle or catheter. However, the diameter of the vessels in small children often does not accommodate the diameter of the curve of the wire tip and so, as the tip of the wire regains its curved shape as it leaves the needle/catheter, it invariably ejects itself from the vessel, resulting in the loss of intravascular access. Interestingly, many small arterial line kits come with the J-wires but we highly recommend straight, soft-tip wires rather than the J-tip wires.

Dilators and catheters

Using a micropuncture needle to access the vessel of interest, an 0.018-inch wire will likely be used to access the vein. Although some 0.018-inch wires have a reasonably stiff shaft, the backbone of the small wire may make dilation of the track and line placement problematic. Therefore the 0.018-inch wire can be exchanged for a larger, 0.035-inch (1 mm) guide wire using a 3–4 dilator (a 3 Fr dilator seated within a 4 Fr dilator). The 3–4 dilator goes over the 0.018-inch wire and then the 3 Fr dilator and wire are removed and the 4 Fr dilator left in place. The 4 Fr dilator will accommodate the 0.035-inch wire. These dilators are standardly included with the micropuncture vascular access kit. A 0.035-inch wire allows for more backbone or body for the easy and safe passage of dilators and lines. Depending on the catheter selected, the surgeon can then place the catheter directly over the wire (Seldinger technique) or place a peel-away introducer through which the catheter can be placed (modified Seldinger technique). In the very small vessels of the neonate, the catheters are made to go over smaller wires, such as 0.018-inch and 0.010-inch (0.25 mm) wires. In these cases, changing wire to 0.035 inch is not helpful or necessary.

Ultrasound

Historically, central venous access was obtained using anatomic landmarks. The introduction of ultrasound for vascular access has made the procedures safer and easier, particularly in children. Although ultrasound guidance is the standard of care in adults and the evidence in pediatrics is quickly mounting, there is a resistance in pediatric surgery to adopt its use widely. This reflects more an inability to use the technology than any real disadvantage to ultrasound. Even the most experienced and technically gifted pediatric surgeons can find ultrasound helpful in gaining access on difficult patients.

In general a high-frequency (10–20 MHz) linear array transducer is used for vascular access. Although higher wavelengths limit penetration depth, this is generally not a problem in vascular access since most targets are superficial and there are great advantages in image quality and special resolution with higher frequencies. It is important to establish clearly left–right orientation on ultrasound prior to attempting access (left side of the probe seen on the left side of the screen is often preferred but regardless should be consistent for the operator). Technical success is also aided by orienting the probe in such a way that the handle does not impede placement of the needle and wire. This is especially true of the hockey stick probe, which has a handle that can be angled away from the working anatomy.

Fluoroscopy

Fluoroscopy is an important tool for CVL placement in the operating room. It can be used to identify key landmarks before attempting access (e.g. the carina in cases of internal jugular [IJ] or subclavian access). It also guides wire advancement and dilator/peel-away sheath placement and ensures appropriate catheter position at the end of the procedure.

POSITION AND PREPARATION OF PATIENT

Position of patient

When using ultrasound, the patient and target area should be placed between the surgeon and the ultrasound machine. A fluoroscopic compatible table should be used with the patient in supine position. The extremity with the target vessel should be extended. If the target is on the trunk, then the arms should be tucked. For jugular and subclavian access, the patient is placed in the Trendelenburg position to distend target veins and prevent air embolism. A shoulder roll can be used to extend the neck and improve ease of access to the IJ. Note that, if a shoulder roll is used, the catheter tip can change position when the roll is removed. Turning the head to the contralateral side is also helpful in IJ and subclavian access. Femoral vein exposure is aided by abduction and external rotation of the leg.

Sterile preparation

Standard skin preparation is used, in most cases consisting of chlorhexidine solution. Hats, masks, and sterile gloves and gowns are worn. If fluoroscopy is to be used, lead should be worn by all those in the operating room. Ultrasound requires a sterile probe cover, with non-sterile ultrasound gel directly on the transducer within the cover and sterile ultrasound gel available for use directly on the patient. Avoiding any air bubbles in the gel layer between the probe and the probe cover is extremely important because air causes shadowing and thus large areas of non-visualization in the ultrasound field of view.

TECHNIQUE

Cut-down technique

Traditionally, central venous access has often been obtained by the cut-down technique by the pediatric surgeon. The vessel of choice is exposed, dissected free, and often ligated proximally and a tie is placed distal around the vessel. A venotomy is then performed and the catheter is carefully inserted through the venotomy site distal to a central location. The catheter is secured by tying down the distal ligature around the vein and the catheter inside the vein. Alternatively, a 7/0 Prolene purse string can be used around the venotomy site in order to avoid ligation of the vessel.

Ideal catheter tip position was discussed above. The cavoatrial junction can be approximated on fluoroscopy as 1.5–2 vertebral bodies inferior to the carina. Therefore, after tunneling to the access site and before placement through the introducer into the IJ, the catheter length is measured. This can be done with either a wire or by laying the catheter on the patient's chest while using fluoroscopy. The catheter is then cut to length and advanced through the introducer. The introducer is removed, and proper position is confirmed with fluoroscopy. Target vessels of such cut-downs include the external jugular vein, facial vein, IJ vein just above the clavicle, and the proximal greater saphenous vein. Overall, the cut-down technique has been largely replaced by percutaneous access. However, the cut-down technique remains an important skill that is indispensable in many circumstances.

Percutaneous technique: Seldinger and modified Seldinger

A Swedish radiologist, Sven-Ivar Seldinger, in 1953 described a novel technique for vascular access involving a needle, a wire, and a catheter. This technique has become the standard for many percutaneous techniques including vascular access.

First, the vessel is accessed with a needle via venipuncture. This is generally best performed using a 21-gauge or 22-gauge micropuncture needle. Using the traditional landmark technique, a syringe is used with mild negative pressure suction applied so that once the vessel lumen is entered, the syringe will have a return of blood. In the authors' experience, when using an ultrasound, a syringe is not used. Reasons for this are as follows:

- The negative pressure can collapse and spasm the very small vessels.
- The process of removing the syringe in small vessels often dislodges the needle from the vessel.
- With correct usage of the ultrasound it is straightforward to determine when the needle tip is in the lumen of the vessel and so blood flash is not needed.

Once the vessel is entered, regardless of technique, a wire is advanced through the needle and into the vessel lumen. When the needle is well within the vessel lumen, the wire is advanced. There should be very little resistance to wire advancement. If resistance is met, ultrasound or fluoroscopy should be used to confirm the position of the wire and guide advancement.

Once the wire is in place, a skin nick with an 11 blade may be helpful adjacent to the wire, avoiding any skin bridge between the two. Dilators, as needed, are passed over the wire to expand the tract to the necessary size, although in the smaller catheters this is unnecessary and often can lead to postoperative oozing. Larger dilators in central vessels should be used with fluoroscopy to ensure the appropriate trajectory of the dilator and avoid vessel injury. Difficulty with dilations can be remedied by switching to a stiffer wire with more backbone, such as an Amplatz Super Stiff™ wire (Boston Scientific, Marlborough, MA, USA). Once the tract is appropriately dilated, the catheter is advanced over the wire.

A modified Seldinger technique can be used with Silastic catheters, given they are usually difficult to place bareback over a wire. In this situation, a combination introducer sheath and dilator are placed over the wire into the vein lumen. The wire and dilator are removed simultaneously, leaving the introducer in place. A finger is quickly placed over the opening of the introducer to prevent back-bleeding and air embolism. The catheter can then be placed through the introducer into the vein lumen and peeled away (split down the middle) and removed.

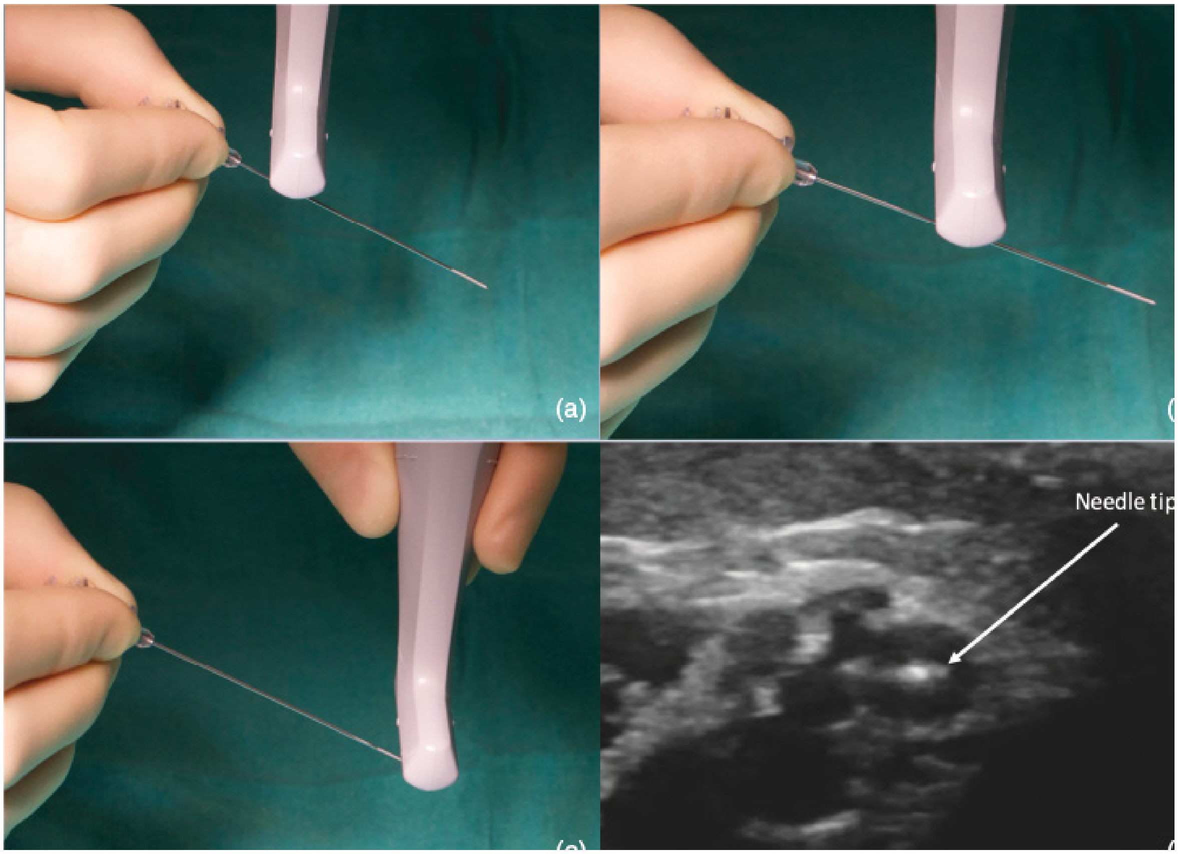
Ultrasound use

Ultrasound can be extremely helpful in vascular access and the American College of Surgeons recommends use of real-time ultrasound for CVC placement. As with any technical skill, however, ultrasound-guided line placement requires appropriate training and practice. With skill development, ultrasound increases the safety, speed, accuracy, and success of CVC placement.

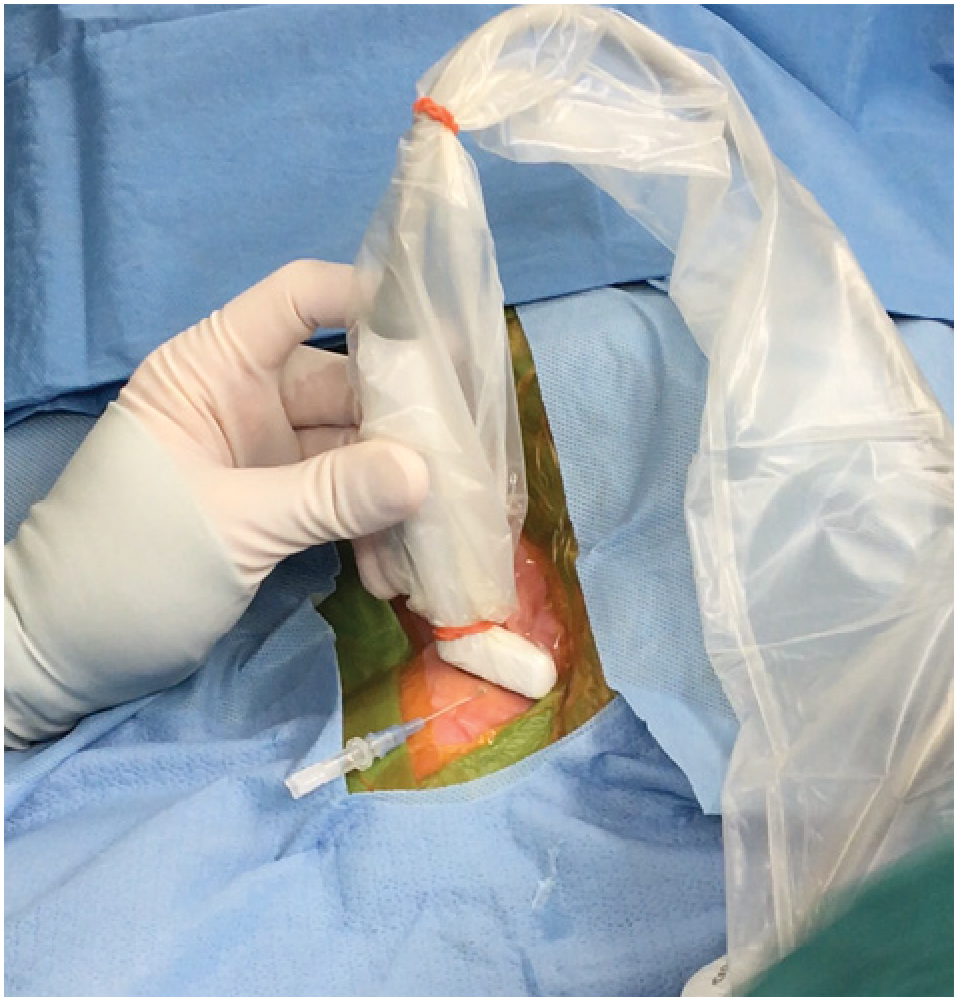
Ultrasound displays three-dimensional (3D) anatomy in two planes. When trying to access a vessel, the operator must therefore choose to view the needle transversely (as a cross-section or circle) or longitudinally (along the length of the needle). The third dimension or *z*-axis can be obtained by moving the transducer perpendicular to its axis.

TRANSVERSE ORIENTATION

The transducer head's long axis is oriented perpendicular to the needle axis, producing a cross-sectional view. The needle is inserted at a 45-degree angle through the skin near the middle of the transducer over the target vessel. The needle is seen in cross-section, and, therefore, appears as only a small point on the display, with the location/depth of the needle tip unknown (**Figure 1.1a–d**). This can be overcome by regularly sliding the transducer down the trajectory of the needle past the needle tip, at which point the bright dot of the needle will disappear. The tip location is confirmed by bringing the transducer back to the needle, resulting in the reappearance of the bright dot on the screen. The needle is then advanced a small distance toward the target vessel and the same technique is repeated to confirm the tip location. In this way the needle is walked down into the vessel. Once the lumen of the vessel is reached, it is important to continue up the lumen of the vessel. The wall of the vessel may spasm at the entry site and the vessel wall can be dragged by the needle for up to 1 cm before the needle actually punctures through. Once the needle has been walked up the vessel lumen, the wire can be placed through the needle and the catheter placed. The transverse orientation is ideal for peripheral IV catheters, PICCs, arterial lines, and femoral lines. Use of the transverse orientation for PICC placement in a neonate is shown in **Figure 1.2**.



1.1a–d With the transverse orientation, the transducer can be anywhere along the length of the the needle (a–c) and still appear as a dot on the ultrasound screen (d).

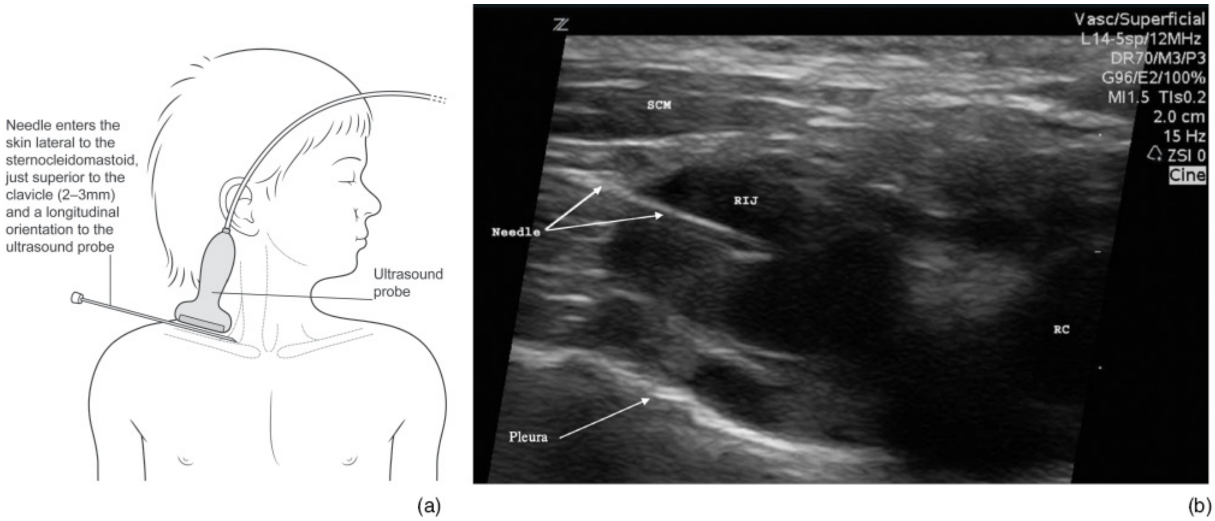


1.2 Example of transverse orientation with the vector of the needle being 90° to the vector of the tranducer.

LONGITUDINAL ORIENTATION

The ultrasound probe is oriented parallel to the needle axis (long axis of needle is directly under the long axis of the transducer) (**Figure 1.3a**). The entire length of the needle is visualized at all times using this method. The needle can then be steered as it is advanced with the tip location known relative to nearby major structures. This orientation is ideal for IJ access, because inadvertent puncture of the carotid artery or lung can be avoided (**Figure 1.3b**). It is sometimes difficult to keep the full needle in view, especially when learning the technique. This leads to the most common problem of losing sight of the needle itself and instead seeing the movement of the surrounding tissue mistaking it for the actual needle.

Losing clear visualization of the needle tip is dangerous and should be avoided. This approach requires patience, because fine adjustments of the transducer head keep it aligned directly over the needle trajectory.


Needle enters the skin lateral to the sternocleidomastoid, just superior to the clavicle (2–3mm) and a longitudinal orientation to the ultrasound probe
Ultrasound probe
(a)
Vasc/Superficial
L14-5sp/12MHz
DR70/M3/P3
G96/E2/100%
MI1.5 TIs0.2
2.0 cm
15 Hz
ZSI 0
Cine
SCM
RIJ
Needle
RC
Pleura
(b)

1.3a,b (a) Orientation of transducer in line with needle. (b) Ultrasound view of needle and key structures when in the longitudinal or in-line orientation (SCM, sternocleidomastoid muscle; RIJV, right internal jugular vein; RC, right carotid artery).

Optimal access to the IJ is obtained using the longitudinal or in-line transducer orientation to allow full visualization of the needle (**Figure 1.3**). The linear, hockey-stick probe (or any suitable-sized linear probe) is placed just superior to the clavicle. The needle enters the skin superior edge of clavicle, just lateral to the sternocleidomastoid muscle (SCM). The visualization from this vantage point is excellent. The distal IJ, subclavian vessels, the carotid artery, and often the brachiocephalic vessels as well as the pleura are all seen from this vantage point. The needle is advanced carefully under complete visualization to the target vessel. With this view, the needle can be seen entering the wall of the vessel and any tenting of the vessel wall by the needle can be seen. Once the needle is in the vessel lumen, the wire is advanced. With this approach, the wire occasionally advances cephalad up the IJ instead of down to the SVC, which can be seen on fluoroscopy. If this occurs, withdraw the wire and under ultrasound visualization turn the needle to point more toward the brachiocephalic vein/SVC. Then replace the wire under fluoroscopy and it will usually go to the SVC.

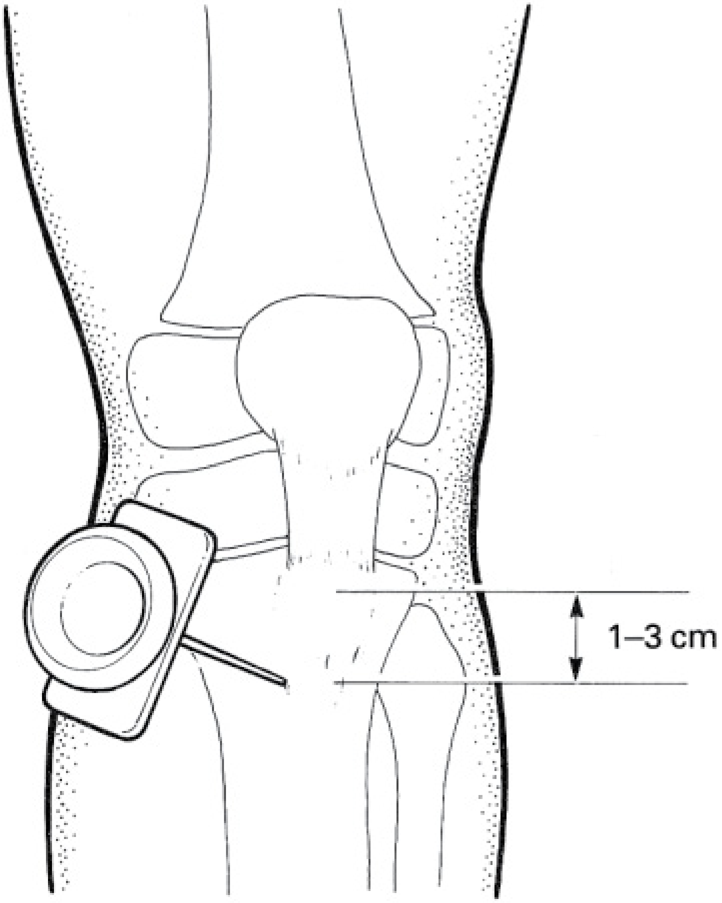
With the wire in place, a subcutaneous tunnel is made from the chest wall incision to the access site in the neck if a tunneled catheter is to be placed. Care must be taken to fashion the tunnel course in a gentle curve, avoiding acute angles, and using the low, lateral neck access site facilitates this.

Tunneling technique

Tunneled IJ venous catheter placement is the procedure of choice when durable, long-term central access is required. A subcutaneous tunnel is created from the access site laterally over the clavicle and inferiorly to the anterior chest to either an external hub or to a subcutaneous port. Accessing the IJ too high on the neck results in a tight loop in the neck. This can lead to kinks in the catheter and displacement of the tip secondary to subcutaneous retraction into the tunnel pocket in the neck. For this reason, the access point should be as close to the clavicle as possible.

EMERGENCY INTRAOSSEOUS VENOUS ACCESS

This route provides immediate vascular access during life-threatening emergencies in young children when rapid venous access cannot be achieved (cardiac arrest, shock, burns, and trauma). Contraindications include fracture or infection near the insertion site. After antiseptic preparation, the skin is punctured with a scalpel blade. The intraosseous needle is inserted into the medullary cavity of the proximal tibia through the middle of its flat anteromedial surface, 1–3 cm below the medial tuberosity (depending on the size of the child) (**Figure 1.4**). The infusion needles (14- to 18-gauge) have an inner occluding stylet designed to facilitate bone penetration and should be inserted almost perpendicularly to the bone, but angled slightly away from the growth plate. Upon entering the marrow cavity, the resistance suddenly decreases. The needle should then stand firmly in the bone. It should be possible to aspirate bone marrow or flush the needle easily without extravasation. The needle flange is adjusted to skin level and taped in position. The patient's leg should be restrained with a support behind the knee. Crystalloids, blood products, and drugs can be infused, but blood sampling may occlude the needle.



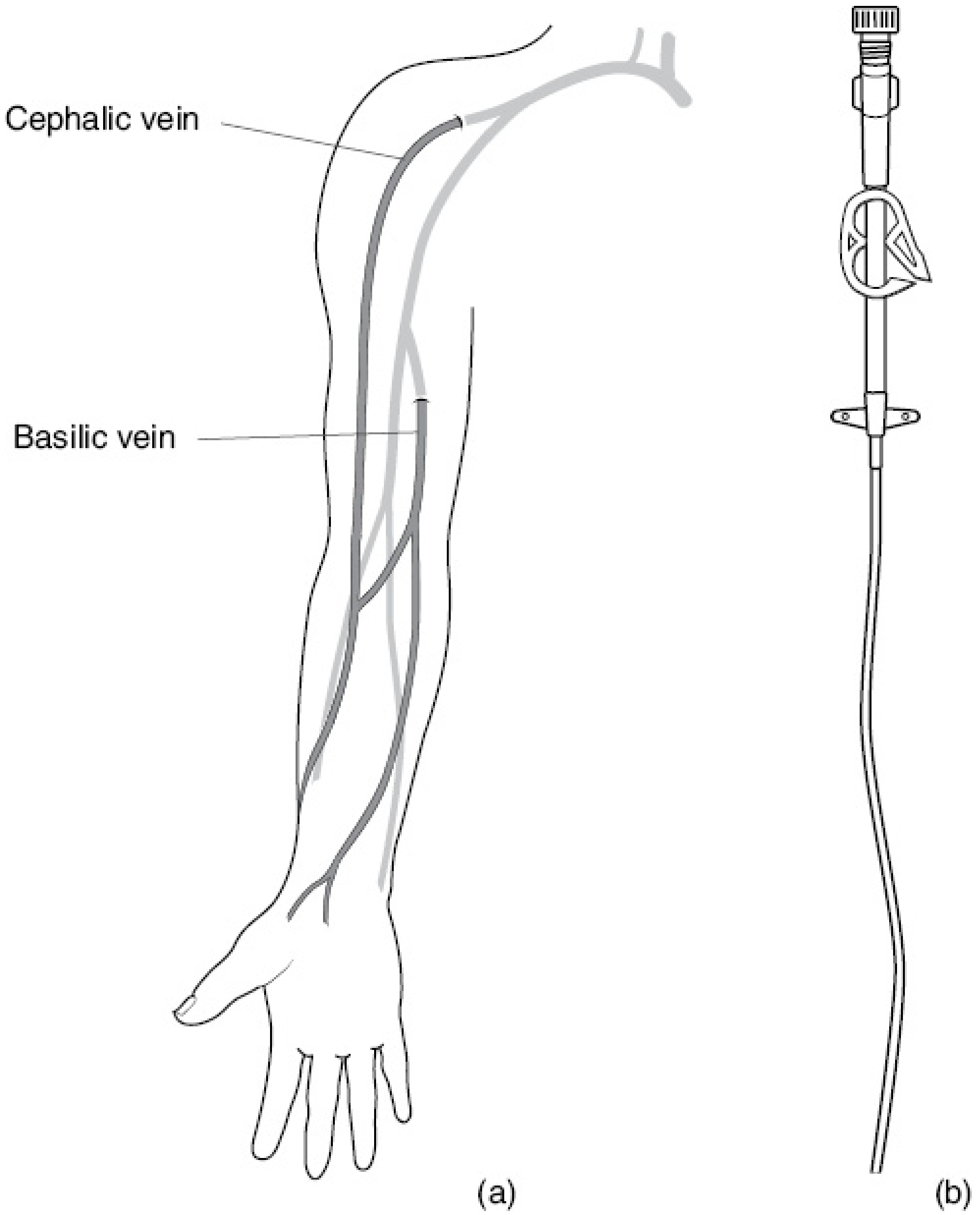
1.4

The infusion needle should be removed once suitable conventional access has been obtained if potential complications (extravasation, compartment syndrome, fractures, osteomyelitis, fat embolism) are to be avoided. The distal femur and distal tibia are alternative sites.

Peripherally inserted central venous catheters

Peripherally inserted central venous catheters (PICCs) are single or dual-lumen catheters typically made of silicone elastomer or polyurethane. They range in size from 2 Fr (0.67 mm) to 6 Fr (2.0 mm). Although they are intended for short- to medium-term use (from a week to a few months), they are occasionally left in for much longer. Upper limb veins are generally used for PICCs, and many are inserted without image guidance. However, imaging with ultrasound can improve success rate and may be required to gain access difficult access patients, and the use of fluoroscopy significantly improves the chance of achieving a suitable final catheter tip position. The ultrasound technique for gaining access is covered under the section "Ultrasound use – Transverse orientation." The procedure is easier to perform in older children with larger cephalic and basilic veins. In certain circumstances when a short duration catheter is required, a device of this type may be used as a tunneled CVC. The advantage over cuffed catheters is that it is easy to remove without sedation or anesthesia.

For peripheral access, the arm is stabilized on a support board. Standard, sterile skin preparation is carried out and a local anesthetic injected after selection of an appropriate entry site to the basilic or cephalic vein, usually above the elbow (**Figure 1.5a and b**). The vein is punctured with a 21-gauge needle or 22-gauge cannula. Aspiration of blood confirms successful puncture. A 0.018-inch (0.5 mm) guidewire is then advanced into the vein. If resistance is felt at this point, the needle or cannula should be repositioned (it is usually too far in). An appropriately sized PICC is selected. The needle or cannula is removed and a peel-away sheath of diameter just sufficient to accept the PICC is advanced over the guidewire. The guidewire should be fixed relative to the patient, and pressure applied over the puncture site as this is done. The guidewire and the dilator of the peel-away sheath are then removed, and the PICC inserted into the sheath. It is usually easier to advance the PICC to a central position if its stiffening wire is left in. In certain places, especially near the termination of the cephalic vein in the deltopectoral groove, it may be easier to fix the stiffening wire and advance the PICC over it, unsupported. When the tip lies in the low SVC or upper right atrium, the peel-away sheath is split and removed, aspiration of blood is confirmed, and the catheter is flushed with normal saline. It is then sutured to the skin, and a transparent occlusive dressing applied. When no suitable superficial vein is available, ultrasound-guided puncture of a brachial vein (vena comitans of the brachial artery) is usually successful.



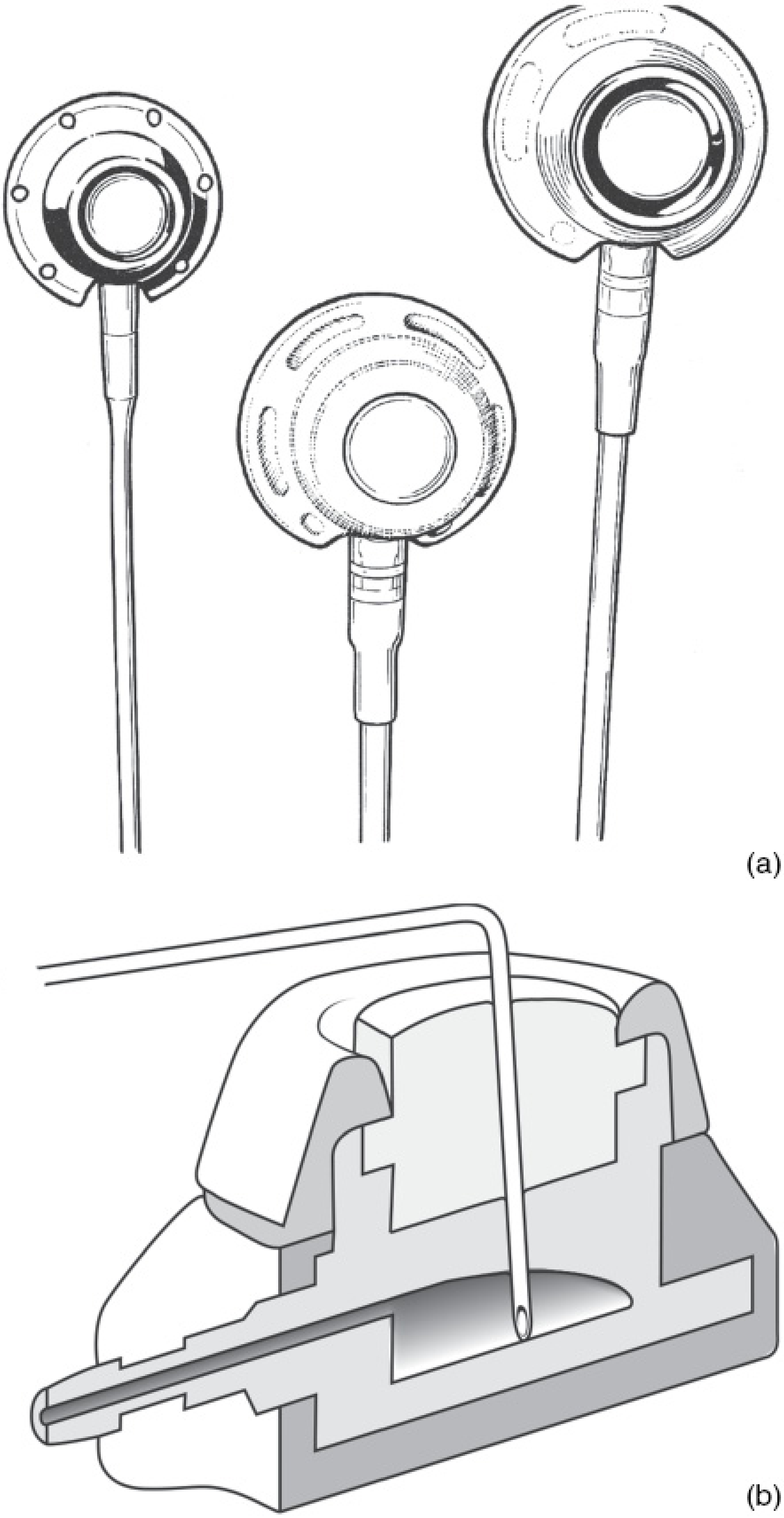
(a)

(b)

1.5a,b

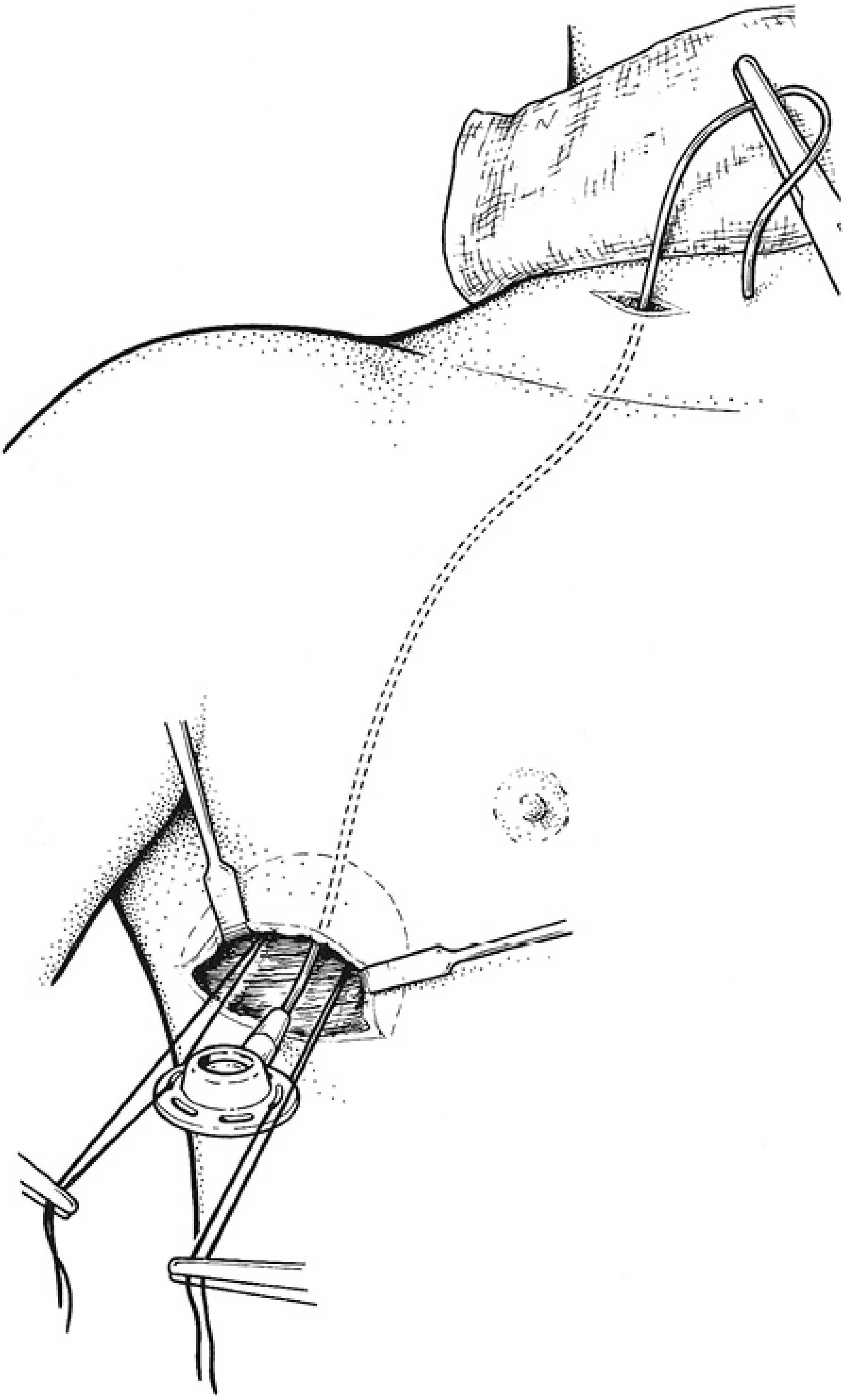
Ports

Totally implantable vascular access devices or “ports” have a catheter connected to a small reservoir, which is implanted subcutaneously (**Figure 1.6a**). A thick silicone membrane forming the roof of the port can be repeatedly injected percutaneously using a 22-gauge side-fenestrated, non-coring (Huber) needle (**Figure 1.6b**). The ports are made from stainless steel, titanium, or hard plastic and are available in different shapes and sizes. Those with a preconnected catheter are easier to insert. One variety is designed to be implanted in the arm with central venous access through a peripherally inserted catheter. Because they have no external catheter, port devices have certain advantages over tunneled CVCs. In particular, they are less likely to require removal for infection, they cannot be accidentally removed, and they allow for activities such as swimming. They are therefore preferable for most children who require only intermittent (e.g. weekly) access, including those with hematological diseases and cystic fibrosis. They are less appropriate in children who cannot tolerate regular needle access, or who require continuous access, for example those who will need intensive chemotherapy or parenteral nutrition.


(a)
(b)

1.6a,b

At the predetermined reservoir site, which must be easily accessible and rest on a firm surface, such as the anterolateral chest wall, the skin incision is deepened with diathermy. Hemostasis must be meticulous. A subcutaneous pocket is developed beneath the superficial fascia in such a way as to avoid placing the port directly under the skin incision. Placement of the port above rather than below the incision may reduce the impact of wound-related problems on port and line function. Before implanting the port, it is helpful to place non-absorbable sutures through the muscular fascia and the circumference of the port; when tied, these provide three-point fixation of the device. The catheter must be tunneled from the reservoir pocket to the site of venous access, such as the internal jugular vein in the neck (**Figure 1.7**). The port is flushed with saline, ensuring there are no kinks in the catheter. The distal catheter is cut to length (see above) and inserted by a cut-down or percutaneous technique. After confirming the catheter tip position by fluoroscopy, the port is flushed with heparinized saline and the skin incision is closed in two layers with an absorbable subcuticular skin suture.



1.7

Each injection must access the port vertically through the center of the silicone diaphragm such that the needle touches the base plate. As the needle is withdrawn, the port should be held in place and positive injection pressure applied to prevent reflux or blood into the catheter. A careful aseptic injection technique must always be used and the system flushed periodically.

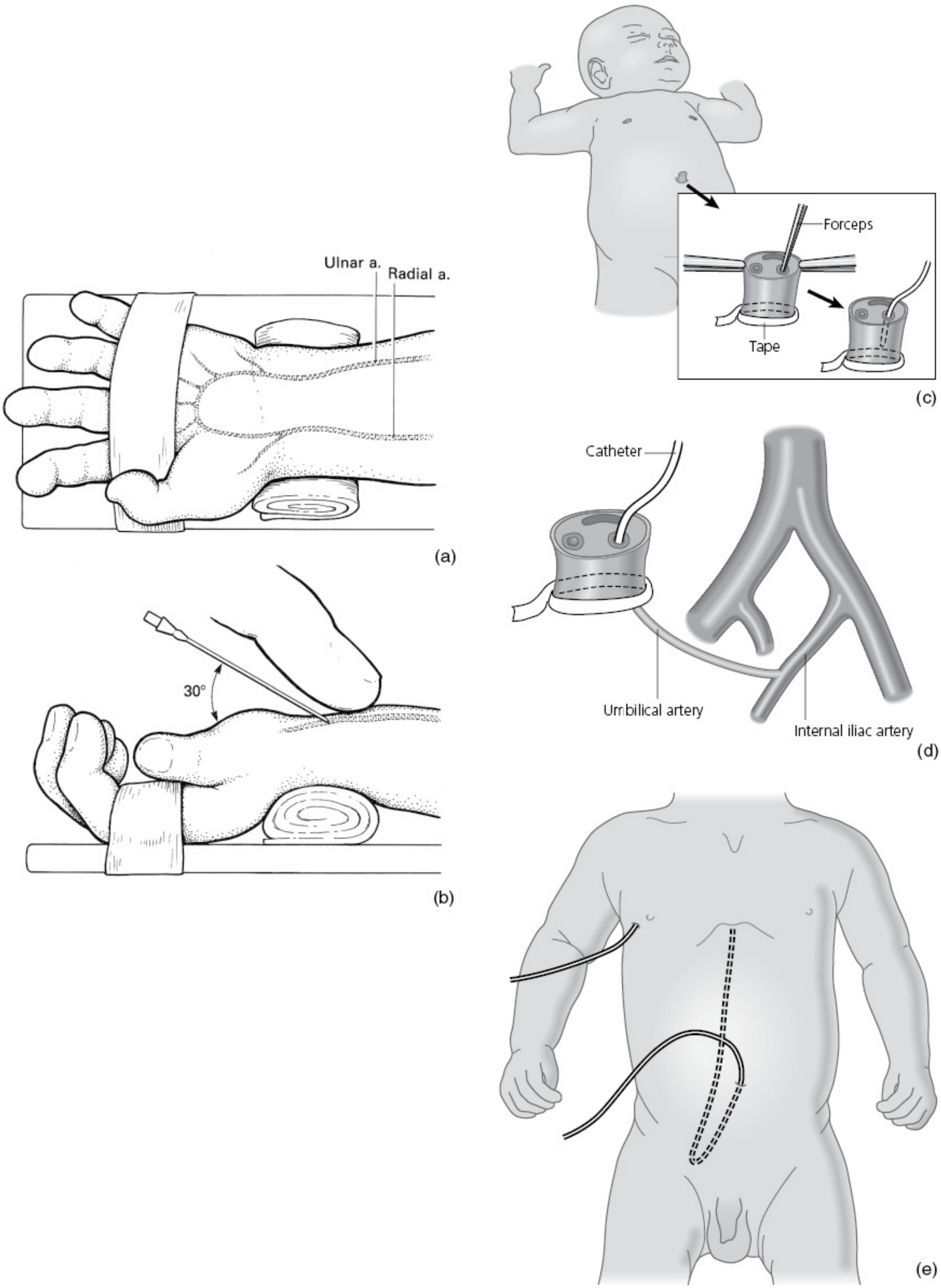
Removal

Most cuffed external catheters and all ports require a short general anesthetic for removal. With the former, the Dacron cuff can usually be dissected free with a hemostat and fine scissors via the exit site incision, which is then closed with absorbable sutures, skin tapes, or biological glue.

ARTERIAL PUNCTURE AND CANNULATION

Intra-arterial access is used to provide continuous monitoring of systemic arterial blood pressure and to enable repeated arterial sampling for blood gas measurements.

The radial artery is the preferred site for both percutaneous and cut-down cannulation (**Figure 1.8a**). The presence of adequate collateral flow must first be checked by the Allen test; both arteries are occluded at the wrist and after releasing the ulnar artery alone, the hand should flush pink (most hands have an ulnar dominant palmar arch). Alternatively, a Doppler ultrasound study may be performed. A small roll is placed under the supinated, extended wrist and the palm is taped to a padded surface, keeping the fingers exposed in order to assess the distal circulation. The skin is cleaned with antiseptic and a small quantity of local anesthetic is injected subcutaneously over the radial artery just proximal to the transverse crease at the wrist. The technique of using ultrasound is described in the section “Ultrasound—Transverse orientation.” If ultrasound or ultrasound skills are not available, percutaneous insertion by palpation can be performed. The artery position is verified by palpation. Two techniques are used. In the first, the needle and Teflon cannula are advanced at about 30 degrees to the skin until a flashback of blood is seen (**Figure 1.8b**). In the transfixion method, the artery is transfixed by the needle and cannula. The needle is then removed and the cannula is gently withdrawn until arterial blood appears when it is advanced up the artery lumen.


Ulnar a.
Radial a.
(a)
30°
(b)
Forceps
Tape
(c)
Catheter
Umbilical artery
Internal iliac artery
(d)
(e)

1.8a–e

In the cut-down technique, a small transverse incision over the artery allows the vessel to be punctured and cannulated under direct vision, with the option of proximal and distal vessel control. The catheter hub is sutured in place and the skin is sutured around the cannula.

Arterial cannulas require continuous perfusion with 0.5–1.0 mL/hour heparinized saline. Because of the risk of serious complications (ischemia, embolism, hemorrhage, sepsis), arterial access requires an even higher level of vigilance and should be used for the shortest possible time. The cannula should be removed if signs of digital ischemia develop. Alternative sites for arterial access include pedal, umbilical, femoral, brachial, and axillary arteries, but complications are more frequent than with radial artery cannulas.

Peripheral artery cannulation is not feasible in some sick infants who require intensive monitoring, and umbilical artery cannulation may be required (**Figure 1.8c and d**). If arterial and venous cannulation is necessary, it is recommended that arterial access (the more difficult) is carried out first. Arterial cannulation is described, but the principles are similar for venous access.

White tape under moderate tension is applied to control the cord at its base. The cord is cut with a no. 15 scalpel about 1 cm from skin level. The two thick-walled umbilical arteries and the one thin-walled vein are identified. A fine non-toothed forceps is introduced closed into the selected artery and opened gently to achieve dilatation. A preprepared and flushed catheter is grasped close to the tip using another pair of forceps and advanced into the umbilical artery, gently but firmly in a caudal direction. The catheter is thereby guided into the aorta via an internal iliac artery. Ready bleeding should be seen and a predetermined position aimed for and confirmed radiologically. In the “low position” the catheter tip is just above the aortic bifurcation, while in the “high position” it is just above the level of the diaphragm (**Figure 1.8e**).

POSTOPERATIVE CARE

The postoperative care of CVCs is crucial to longevity and optimal function. Tunneled CVCs should be looped then secured with occlusive, transparent dressing. This ensures that distal traction is not directly transmitted to the line at the exit site. The development of dedicated teams and carefully defined protocols that cover aspects of management, such as frequency of flushing and line handling techniques for sampling and infusions, has also helped to reduce complications. If well cared for, the majority of catheters last the duration of the planned course of treatment. However, complications which may shorten this duration are well recognized and are summarized in **Table 1.2**.

Table 1.2 Complications of central venous access

Timeline	Complication	Prevention	Management
Immediate	Pneumothorax	Avoid blind procedures	Chest drain
	Hemorrhage	Meticulous technique	Digital pressure, vein repair
	Air embolism	Positive pressure ventilation	Patient positioning, resuscitation
	Arterial puncture	Avoid blind procedures	Digital pressure
	Cardiac arrhythmia	Fix guidewire	Remove irritation, carotid massage, adenosine
	Malposition	Intraoperative fluoroscopy	Line revision
Early	Accidental removal	Secure exit site suture, loop catheter	Line replacement
	Catheter-related sepsis	Meticulous aseptic technique for insertion and subsequent line access	Antibiotic therapy, catheter removal
	Chest wall hematoma	Check and normalize coagulation parameters preoperatively	Pressure dressing
	Cardiac tamponade	Site stiff percutaneous catheters in low SVC	Pericardiocentesis, pericardiotomy
Delayed	Catheter-related thrombosis	Site catheter tip in upper right atrium, flush regularly	Conservative management, anticoagulation, thrombolysis
	Extravasation		
	Port–catheter separation	Use of preconnected catheter	Urgent exchange
	Catheter migration	Site catheter tip in upper right atrium	Catheter revision
	Catheter fracture	Gentle handling and careful removal	Fluoroscopic catheter retrieval of embolized fragments

FURTHER READING

Arul GS, Lewis N, Bromley P, Bennett J. Ultrasound-guided percutaneous insertion of Hickman lines in children: Prospective study of 500 consecutive procedures. *J Pediatr Surg* 2009; 44: 1371–6.

Brevetti LS, Kalliainen L, Kimura K. A surgical technique that allows reuse of an existing venotomy site for multiple central venous catheterizations. *J Pediatr Surg* 1996; 31: 939–40.

Bruzoni M, Slater BJ, Wall J, et al. A prospective randomized trial of ultrasound- vs landmark-guided central venous access in the pediatric population. *J Am Coll Surg* 2013; 216(5): 939–43.

Criss CN, Gadepalli SK, Matusko N, Jarboe MD. Ultrasound guidance improves safety and efficiency of central line placements. *J Pediatr Surg* 2019; 54(8): 1675–9.

Donaldson JS, Morello FP, Junewick JJ, et al. Peripherally inserted central venous catheters: US-guided vascular access in pediatric patients. *Radiology* 1995; 197: 542–4.

Hind D, Calvert N, McWilliams R, et al. Ultrasonic locating devices for central venous cannulation: Meta-analysis. *BMJ* 2003; 327(7411): 361.

National Institute for Health and Clinical Excellence, UK (NICE). *Guidance on the use of ultrasound locating devices for placing central venous catheters*. Technology Appraisal Guidance, No. 49. London: NICE, September 2002.

Sigaut S, Skhiri A, Stany I, et al. Ultrasound guided internal jugular vein access in children and infant: A meta-analysis of published studies. *Paediatric Anaesth* 2009; 19(12): 1199–206.

Stringer MD, Brereton RJ, Wright VM. Performance of percutaneous silastic central venous feeding catheters in surgical neonates. *Ped Surg Int* 1992; 7: 79–81.

Vo JN, Hoffer FA, Shaw DW. Techniques in vascular and interventional radiology: Pediatric central venous access. *Tech Vasc Intervent Radiol* 2010; 13: 250–7.

PART I

Head and neck

2

Head and neck surgery

JEFFREY W. GANDER

PREAURICULAR SINUS

PRINCIPLES AND JUSTIFICATION

The preauricular sinus (PAS) is a congenital sinus whose opening is situated on the anterior aspect of the helix of the pinna. It results from ectodermal inclusions that arise during fusion of the six fetal tubercles that form the pinna. The course of the sinus is unpredictable and often consists of multiple branches. It passes anteriorly and inferiorly through the subcutaneous tissue to end in a group of preauricular cysts and frequently extends deep to the subcutaneous tissue to be attached to the cartilage of the pinna. The sinus is often familial and may be unilateral or bilateral. In many cases, it is completely asymptomatic and requires no treatment. However, it may present with an intermittent discharge or become infected. Total excision of the PAS and the underlying group of preauricular cysts is essential for cure of the symptomatic sinus. This may also require excision of a small amount of underlying cartilage at the point of attachment. Incomplete excision results in a significant recurrence rate.

PREOPERATIVE

Definitive surgery should not be undertaken in the presence of active infection, which should be treated with anti-staphylococcal antibiotics. In the case of abscess formation, this requires either aspiration or incision and drainage. At the time of formal excision of the sinus, prophylactic antibiotics should be administered in those cases that have had previous episodes of infections.

The decision about treatment is made on clinical grounds. No further investigations are routinely required preoperatively.

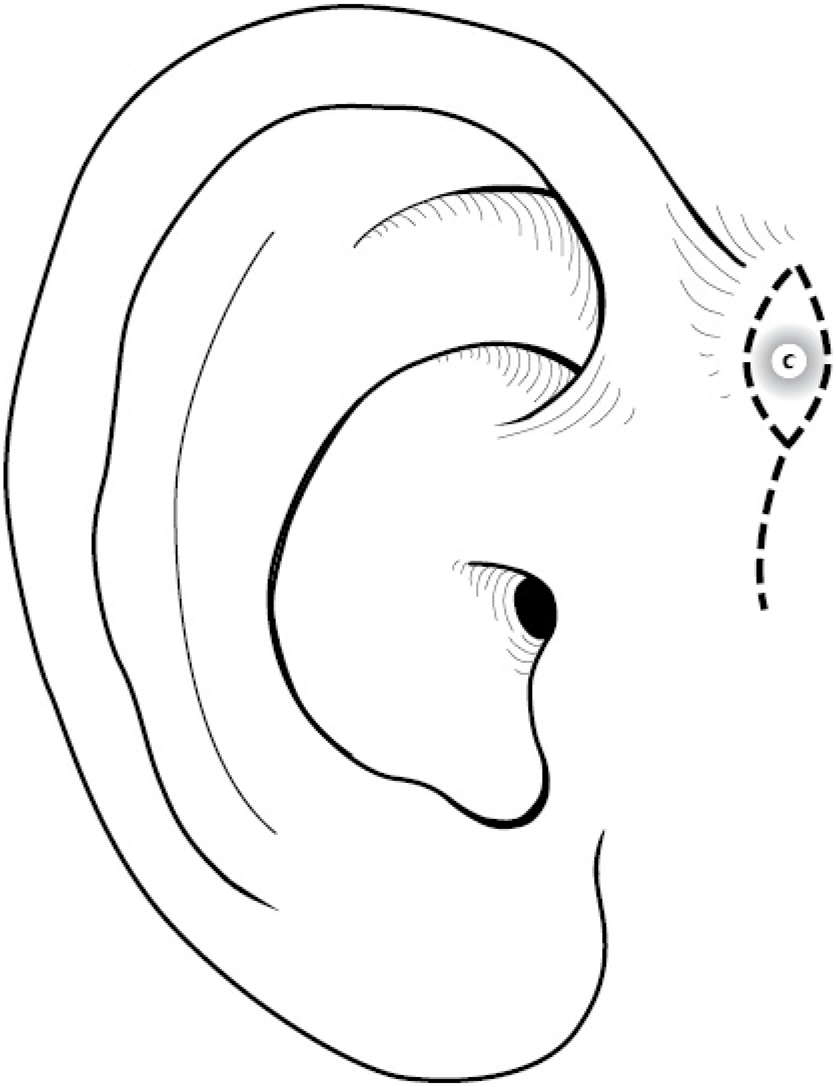
OPERATION

Skin preparation and draping

The child is placed supine on the operating table and the head positioned on a head ring. Any hair anterior to the pinna should be shaved. If bilateral sinuses are present, a head towel is used to maintain a sterile field. In the case of a unilateral sinus, a standard four-drape technique can be used. The injection of methylene blue has been advocated as a method for identifying the ramifications of the sinus. While some surgeons use this routinely, many find that in practice it does not prove helpful.

Incision

An elliptical incision is made around the punctum (**Figure 2.1**). The ellipse should also include any scarring that has resulted from previous infection. The incision is extended inferiorly in a vertical plane immediately in front of the pinna. An alternative incision is an inverted L-shape with an associated skin flap.



2.1

Dissection and excision

The ellipse of skin containing the punctum and underlying sinus is then held using either toothed Adson forceps or a hemostat. Dissection is continued and the underlying group of preauricular cysts identified. Sometimes, insertion of a lacrimal probe is helpful to identify the course of the sinus.

Further dissection is performed ensuring that the skin ellipse, sinus, and cyst are dissected en bloc. As stated above, the deep attachment of the sinus is often lying on the cartilage, and excision of a small amount of cartilage at the point of attachment is often required to prevent recurrence. During deep dissection, particular care is taken to preserve the superficial temporal artery and the preauricular nerve. The sinus, together with the collection of cysts, is then removed and sent for routine histopathology.

Closure

Interrupted fine absorbable sutures are used to close any defect in the cartilage. Drainage of the wound is not routinely required. Bupivacaine 0.25% is infiltrated into the wound to ensure postoperative analgesia. The remainder of the wound is closed in the typical fashion with absorbable sutures.

COMPLICATIONS

- *Recurrence*: 2–50%
- *Wound infection*: 0–30%

OUTCOME

Postoperative hematoma and infection can occur and incomplete excision may result in recurrence of the sinus. This risk is increased in those cases in which recurrent infection has preceded excision, as the dissection planes are often less clearly defined.

EXTERNAL ANGULAR DERMOID CYST

PRINCIPLES AND JUSTIFICATION

Dermoid cysts are congenital cysts that result from sequestration of ectodermal and mesodermal elements. They occur along lines of embryologic closure, with the external angle of the supraorbital ridge being the most common site. External angular dermoid cysts are located underneath the muscular layer and lie in a shallow depression in the outer table of the bone of the skull. A "pit" within the bone is invariably present in the base of the bony depression through which the cyst receives its blood supply. Occasionally, these cysts have an intracranial extension in a "dumb-bell" fashion, although this is far less common than with dermoid cysts situated at the internal angle of the eye or over the bridge of the nose.

External angular dermoids present as rounded, soft, semi-mobile swellings in the lateral portion of the eyebrow. They are usually asymptomatic, although with time can enlarge and rupture or become infected. Treatment is by surgical excision of the cyst, which must be complete to prevent recurrence.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

The diagnosis is made on clinical grounds. If the cyst is in the typical position, near the lateral portion of the eyebrow, no imaging is needed. If it is located near the medial canthus of the eye, along the nose or a cranial suture line, a magnetic resonance imaging (MRI) or computed tomography (CT) scan should be performed to look for intracranial extension. Skull X-rays are typically not helpful for excluding an intracranial extension.

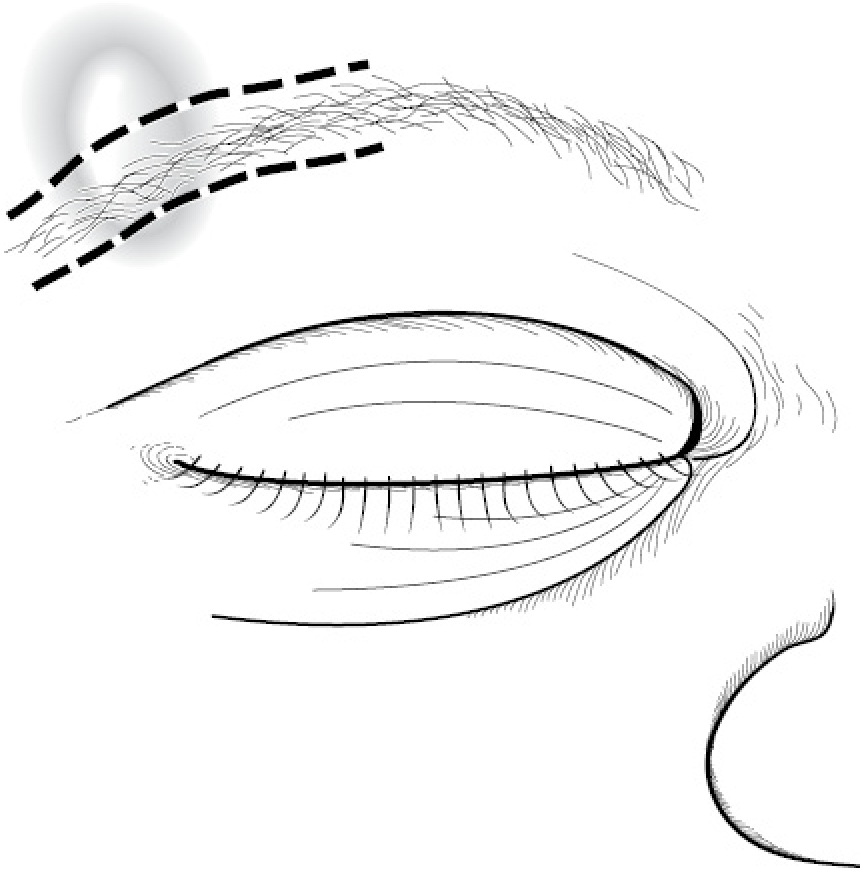
OPERATION

Skin preparation and draping

There is no need to shave the eyebrow for the procedure. The eyelids are taped closed or covered with a plastic adhesive to prevent soiling of the eye. Scalp hair can be matted down with a water-based lubricant jelly to avoid it coming into the field. A head towel can be used to maintain the sterile field.

Incision

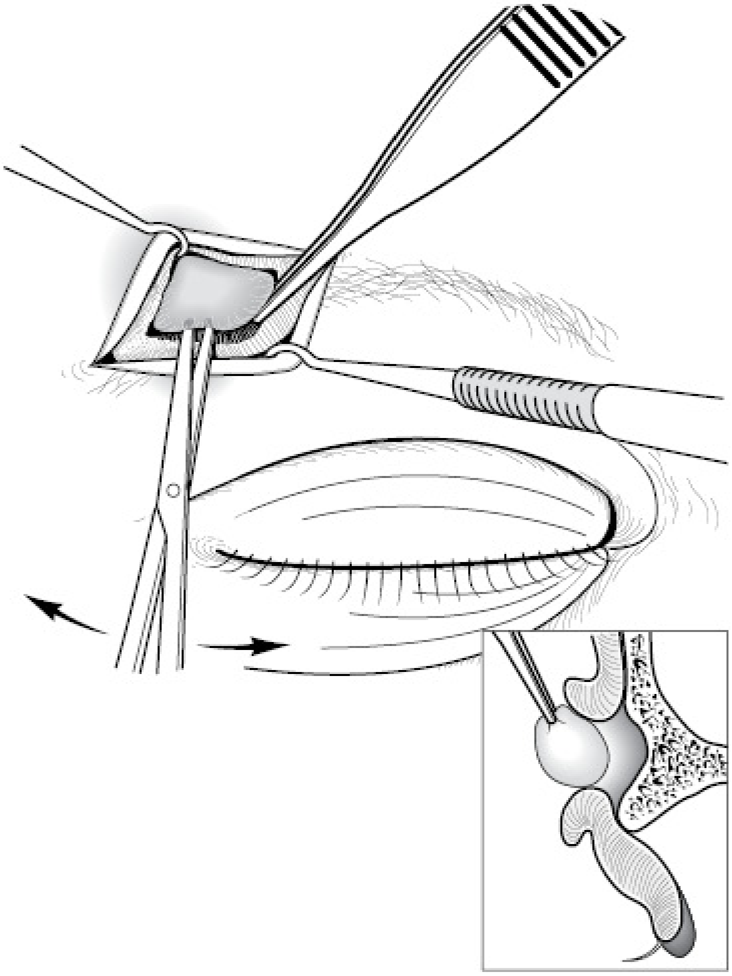
A 1.0–1.5 cm incision is made over the cyst in line with the upper or lower margin of the eyebrow (**Figure 2.2**). Care is taken not to cut the hair follicles of the eyebrow. Although this incision gives a good cosmetic result, an alternative approach is to make the incision in the superior eyelid crease.



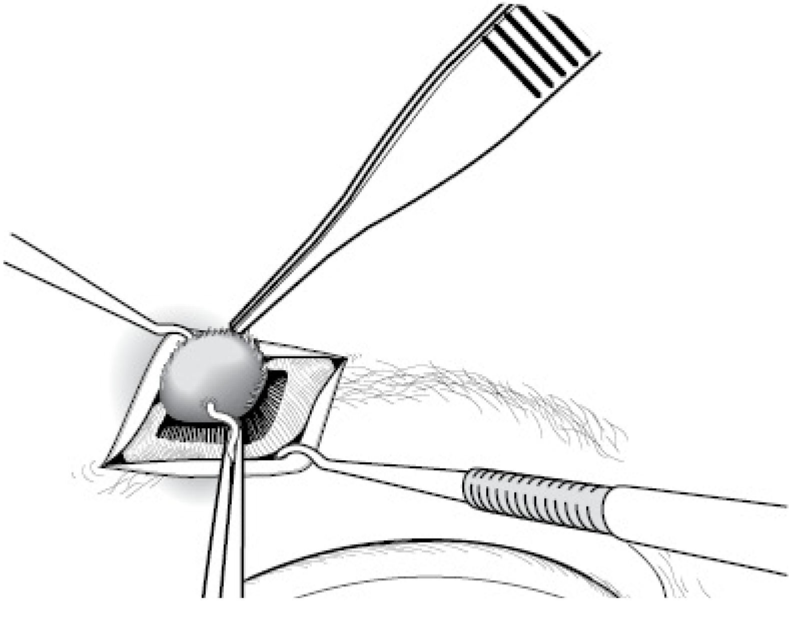
2.2

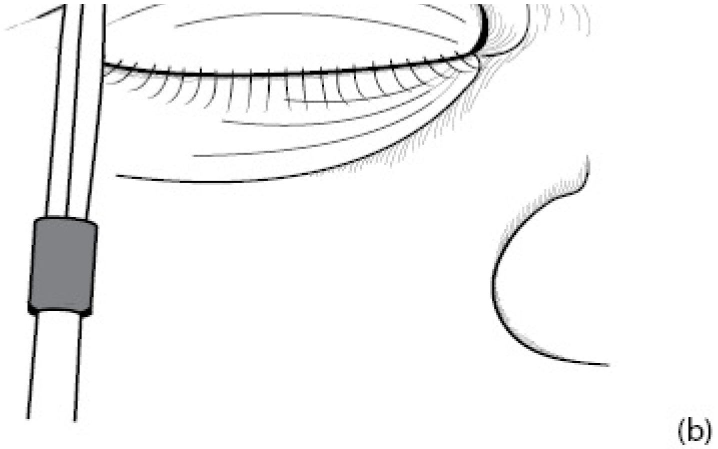
Dissection and excision

The cyst is then mobilized using fine haemostats (**Figure 2.3a**). Instruments should not be applied directly to the cyst as this may result in its rupture. The cyst is freed from its deep attachments, which can require excising a circumferential rim of periosteum in continuity with the cyst (**Figure 2.3b**). If present, the feeding vessels passing through the pit of the bony depression are coagulated. Occasionally, the pit needs to be packed with bone wax to secure hemostasis. If rupture does occur during mobilization, the wound is irrigated with saline and the contents and lining of the cyst should be completely removed to prevent recurrence.



(a)





2.3a,b

MINIMALLY INVASIVE APPROACHES

While both of the incisions described above usually give good cosmetic results, endoscopic-assisted procedures have been described that aim to make any scarring even less conspicuous. Two to three ports are used and placed in the hairline. Insufflation of a pressure of 4 mmHg with CO_2 is used and the dissection performed. Concerns over this technique of facial nerve injury and incomplete excision have been raised. Additionally, it can be technically demanding as well as a higher cost than direct excision.

In the rare event of an intracranial extension, a formal craniotomy is required because the intracranial portion may be the larger of the two elements.

OUTCOME

Complications are rare following this procedure. Postoperative hematoma and infection can occur, and incomplete excision may result in recurrence of the cyst.

BRANCHIAL CYSTS, SINUSES, AND FISTULAS

HISTORY

The branchial (external) and pharyngeal (internal) arches are structures which recapitulate our ontogeny with perhaps four or five being identifiable. In fish and most amphibians these become functional gill slits. The word itself comes from the Greek word for gills (βράγχια). Ascherson in 1832 was the first to suggest that the lateral cervical cysts he observed derived from such embryological precursors, using the term branchial cysts.

PRINCIPLES AND JUSTIFICATION

Cysts, sinuses, and fistulas of the neck derived from branchial cleft remnants are relatively common in children (Table 2.1). Although all are present at birth, sinuses and fistulas are encountered more commonly in infants and young children, while cysts present more often in older children and adolescents. Remnants of the first and second branchial apparatus are most common, accounting for more than 95% of all branchial anomalies, with abnormalities of the second cleft outnumbering those of the first by about 6 : 1. Abnormalities of the third and fourth branchial apparatus are rare, but case reports and reviews indicate that they may be more common than previously thought.

Table 2.1 **Derivatives from branchial apparatus**

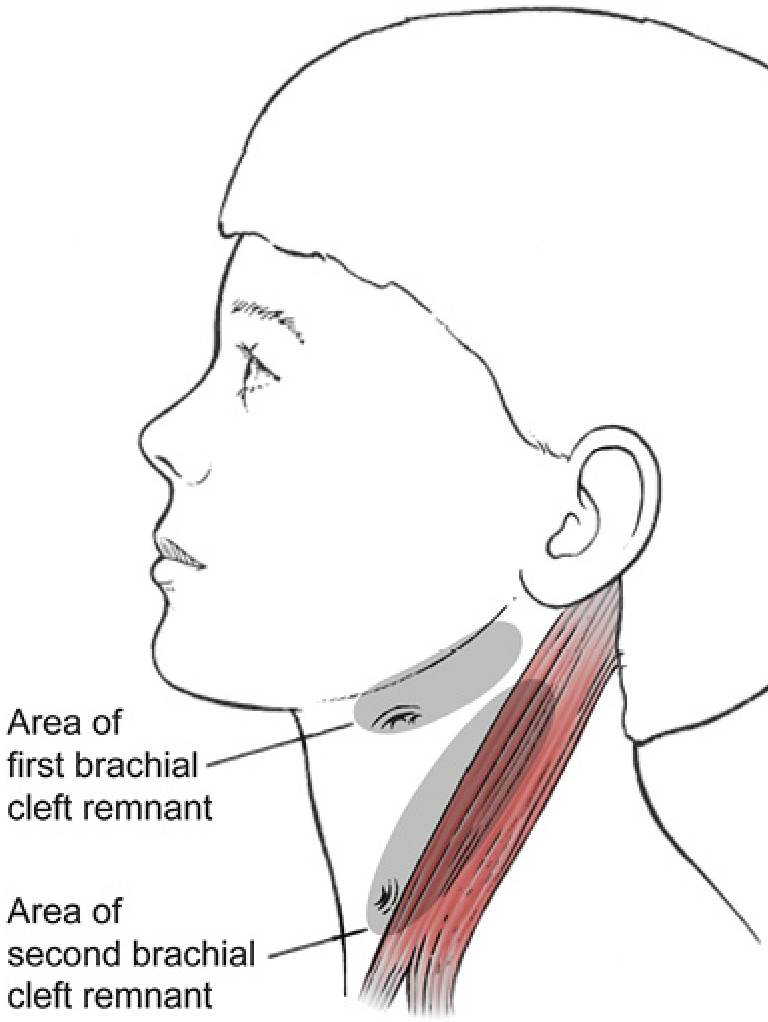
Arch/Pouch	Normal structures	Clinical correlate	Surface marking
I	Eustachian tube, middle ear Mandible Incus, malleus	Cyst/sinus/fistula	Line from external auditory meatus toward angle of jaw
II	Hyoid Stapes	Cyst/sinus/fistula	Just anterior to SCM (cyst) Distal SCM (fistula)
III	Hyoid Inferior parathyroids Thymus	Thyroiditis	
IV	Laryngeal cartilages Superior parathyroids C cells of thyroid	Thyroiditis	

Knowledge of head and neck embryology is helpful in understanding these abnormalities. The branchial arches appear by the 15th day of embryonic life and present as bar-like ridges separated by grooves or clefts. Five paired ectodermal clefts and five endodermal pouches separate the six branchial arches. A closing membrane lies at the interface of the pouches and clefts.

The pathogenesis of branchial cleft anomalies is controversial, and may occur as any combination of sinus, fistula, and cyst. Incomplete obliteration of the branchial apparatus, primarily the cleft, is accepted as the most likely etiology. Most branchial anomalies arise from the second branchial apparatus as the second branchial arch overgrows the second, third, and fourth branchial clefts, and finally fuses with the lateral branchial wall. As the arches coalesce during the growth of the embryo, part of the first branchial cleft remains open as the Eustachian tube and auditory canal. The second branchial cleft normally closes completely; however, either branchial cleft may form a sinus tract or cyst as it coalesces.

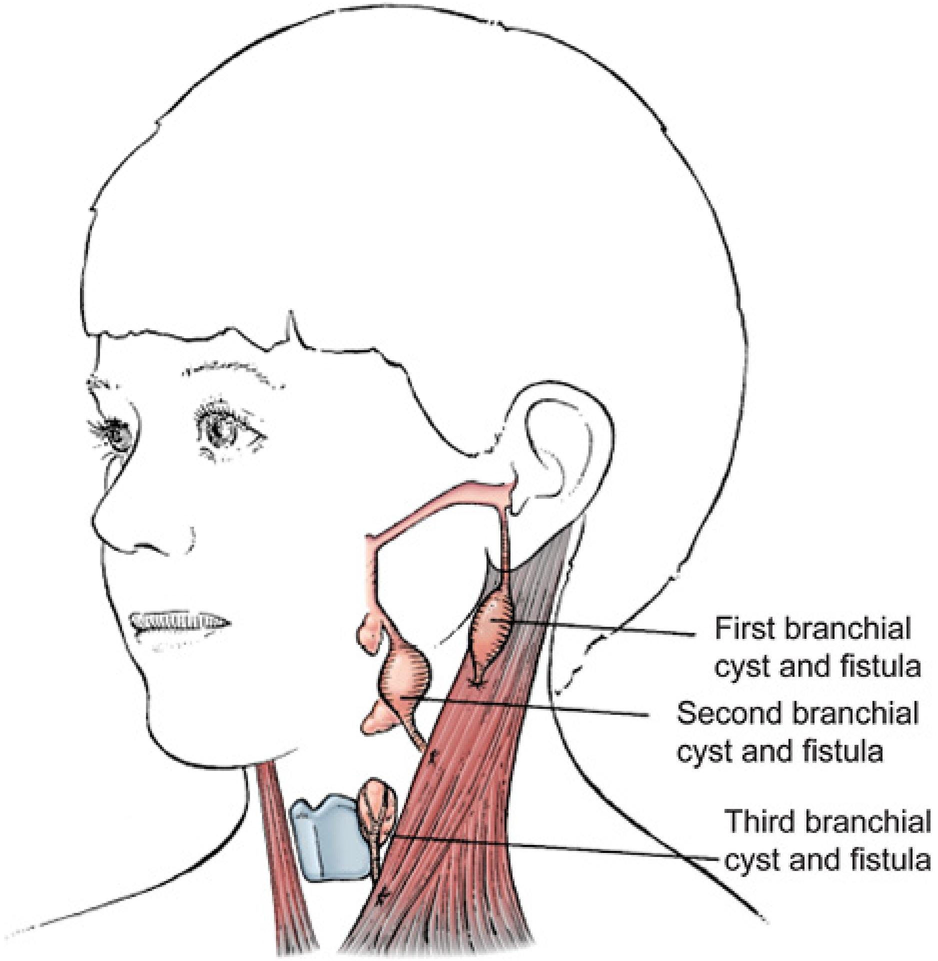
Although branchial apparatus anomalies may present at any age, most branchial sinuses present clinically soon after birth or before the age of ten years.

Remnants of the first branchial cleft, usually a short sinus track occasionally opening into the external auditory canal, occur along a line extending from the auditory canal behind and below the angle of the mandible to its midpoint. Second branchial cleft remnants are found anywhere along a line extending from the tonsillar fossa down to a point on the lower third of the anterior border of the sternocleidomastoid muscle (SCM) (**Figure 2.4**). The more common second branchial cleft sinus presents as a pinpoint opening on the anterior border of the SCM, just short of its sternal origin. The defect is usually characterized by the appearance of small drops of clear fluid at the opening or by the occurrence of infection in the tract itself. The anomaly may be either unilateral or bilateral and may be familial. Tracts that have an exterior opening occasionally become infected, although infection is a more common problem in sinuses and cysts in the older age group.



2.4

Cysts of the first branchial cleft usually present as enlarging masses near the lower pole of the parotid gland and are more commonly seen in older children and young adults. Cysts of the second branchial cleft usually present in children and young adults as a mass at the mandibular angle along the anterior border of the SCM, often associated with upper respiratory infection (**Figure 2.5**). Case reports and reviews suggest that cysts or acute infections arising from a third or fourth branchial pouch sinus are rare but well-defined entities that offer diagnostic and therapeutic challenges not encountered with the anomalies of the first and second branchial remnants. These lesions present as an air-containing inflammatory lateral neck mass, often posterior to the SCM, in the neonate or as acute suppurative thyroiditis in the infant or child. The etiology for both presentations is a fistulous track from the piriform sinus, most commonly on the left side, occurring as a result of a persistent remnant from the third or fourth branchial pouch. This condition should always be suspected in a neonate presenting with an inflammatory lesion containing air in the left side of the neck. Similarly, acute suppurative thyroiditis is rare and its presence should prompt consideration of a piriform sinus fistula as the etiology. Treatment of the acute infection should be followed by surgical extirpation in all cases.


First branchial cyst and fistula
Second branchial cyst and fistula
Third branchial cyst and fistula

2.5

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

Sinuses of the first and second branchial clefts are diagnosed by their clinical appearance. No special diagnostic imaging is indicated. Ultrasound can be useful with diagnosing cysts. Timing of the operation is considered elective, but the author generally waits until the patient is 9–12 months of age; however, some argue to wait until 2–3 years for concerns of anesthetic on the developing brain. If infection is present, a course of antibiotics should be administered first. If this fails, then incision and drainage or aspiration alone is indicated.

With respect to cysts or sinuses suspected to be of third or fourth branchial pouch origin, a contrast esophagram may demonstrate the presence of a piriform sinus fistula, particularly after a course of antibiotics and resolution of the surrounding inflammation. CT scan has also proved useful in diagnosing such lesions. If imaging techniques are not successful after resolution of the inflammation, then the next time the inflammation recurs, compression of the pus-filled cyst during endoscopy may reveal the origin of the fistula as pus exudes from the piriform sinus.

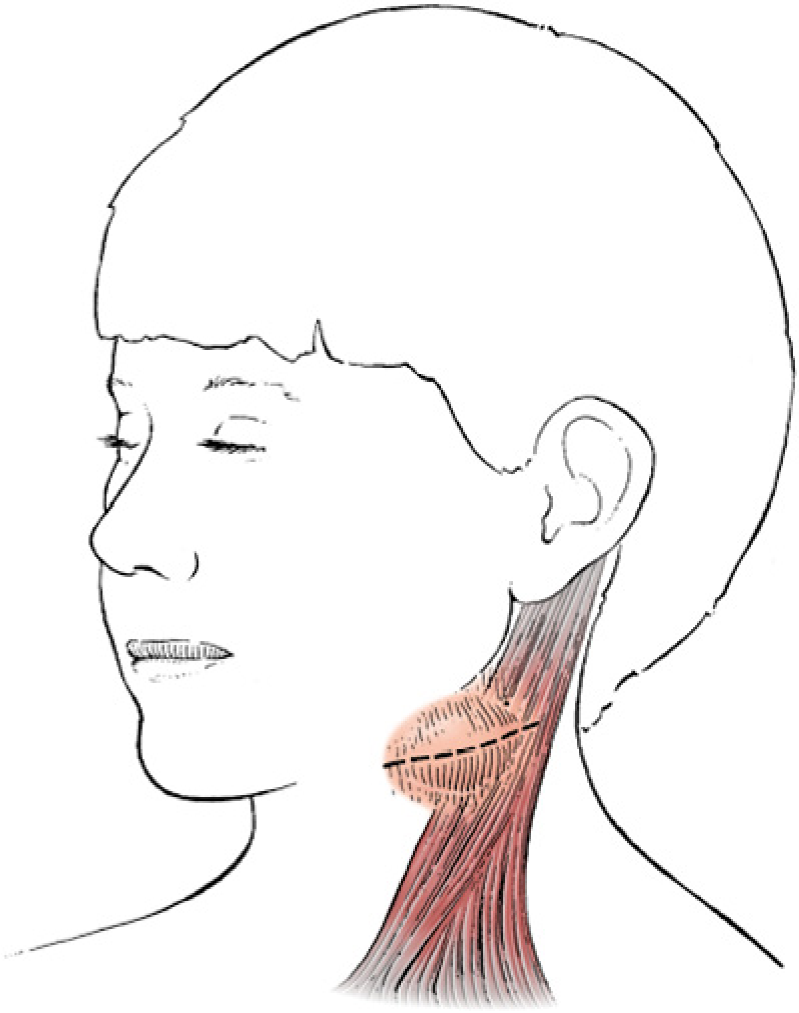
OPERATIONS

Position of patient and preparation

The child is placed in the supine position with a roll beneath the shoulders and a soft ring headrest beneath the cranium so that the neck is extended. Occasionally, tape needs to be applied to the chin to extend the short neck of an infant. The chin is turned away from the side of the lesion.

Exposure of cyst

The most common is derived from the second branchial cleft. The skin incision is made over the cyst in a natural skin crease in order to obtain the best cosmetic result (**Figure 2.6**).



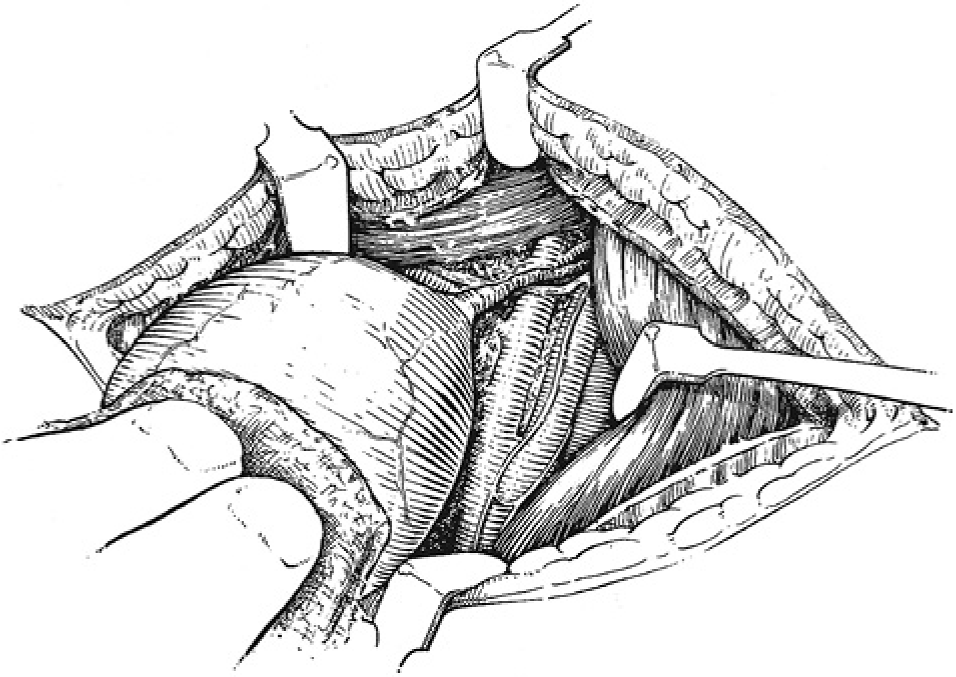
2.6

The incision is carried through the subcutaneous tissues and platysma to the level of the cyst. The cyst is exposed by retracting the skin and muscle flaps and is best accomplished with a self-retaining or ring retractor.

The deep cervical fascia is divided next to the anterior border of the SCM, allowing the belly of the muscle to be retracted away from the cyst. Exposure is extended anteriorly and medially by retraction of the sternohyoid muscle.

Dissection and removal of a second branchial cleft cyst

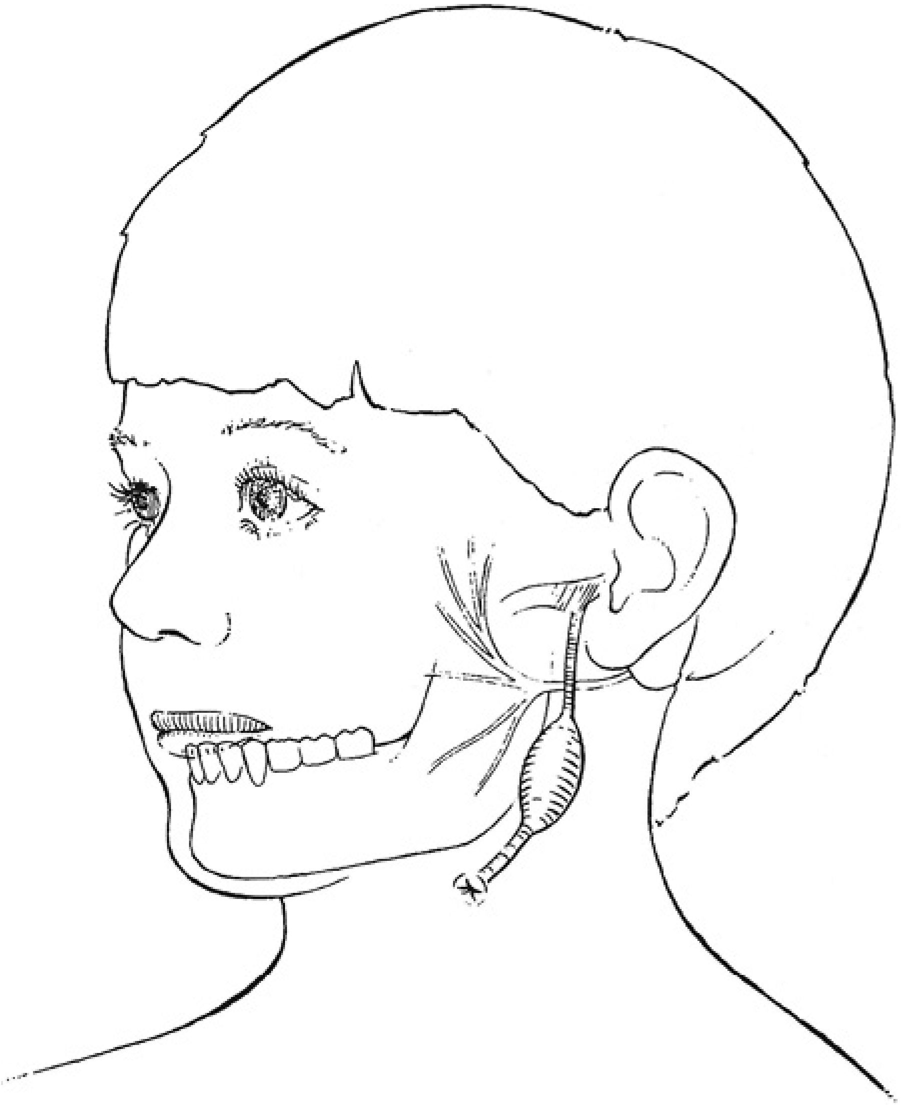
Care is taken to avoid rupture of the cyst, as a tense cyst wall is easier to define and dissect than a collapsed cyst. Adjacent structures are separated from the cyst with special care being taken along the deep aspect where the jugular vein and carotid arteries are in intimate relation. The pedicle of the cyst generally lies posterior to the jugular vein, usually coursing between the carotid artery bifurcation (Figure 2.7). It is then dissected cephalad toward the tonsillar pillar, where it is clamped and suture ligated with a fine absorbable suture.



2.7

Dissection and removal of a first branchial cleft cyst

During dissection of the first branchial cleft cyst, care must be taken to avoid damage to the adjacent facial nerve in cases where there is a tract leading up to or into the external auditory meatus (Figure 2.8). The deep or superficial lobes of the parotid gland may need to be mobilized or occasionally the superficial lobe may need to be resected if the tract extends deeply. A neurosurgical nerve stimulator is often helpful during the dissection. Tracts that extend toward the external auditory canal may require removal of the skin and cartilage components of it.

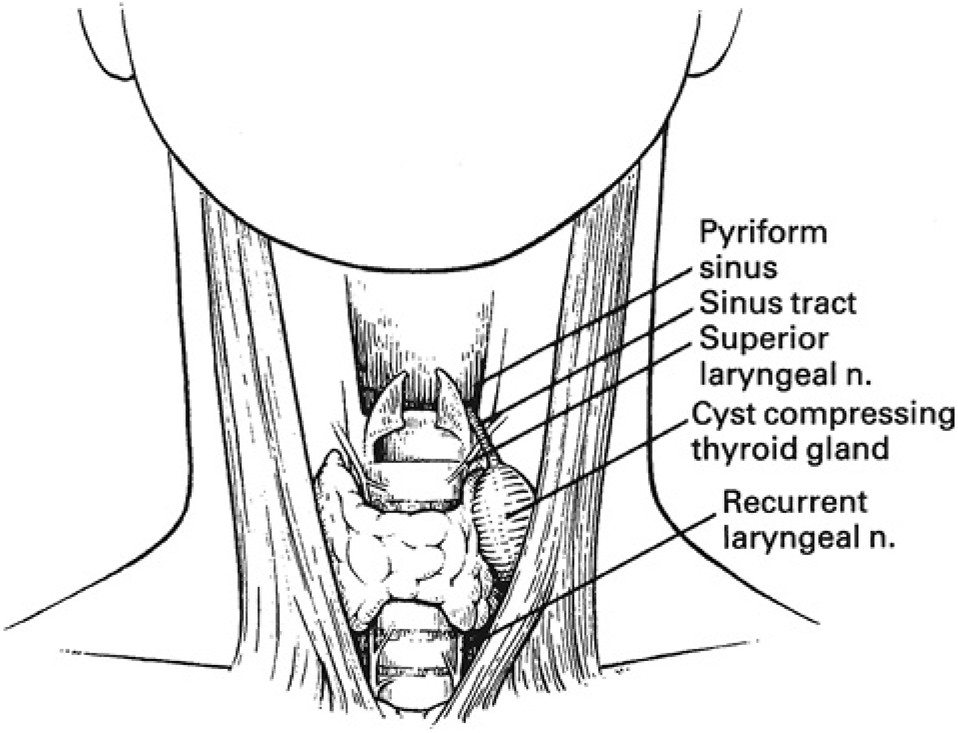


2.8

Dissection and removal of a third or fourth branchial pouch cyst and sinus

Cysts and sinuses of the third and fourth branchial pouch are clinically similar because of their common origin in the piriform fossa and presentation as a neck or thyroid abscess. Exploration of the neck with excision of the entire tract to the level of the piriform sinus is necessary to prevent recurrence. Operative endoscopy at the start of the operation may enable cannulation of the tract from above, which greatly facilitates localization of the tract during resection. Once the cyst and tract are resected, the histological finding of squamous cell epithelial lining confirms the diagnosis of a branchial anomaly.

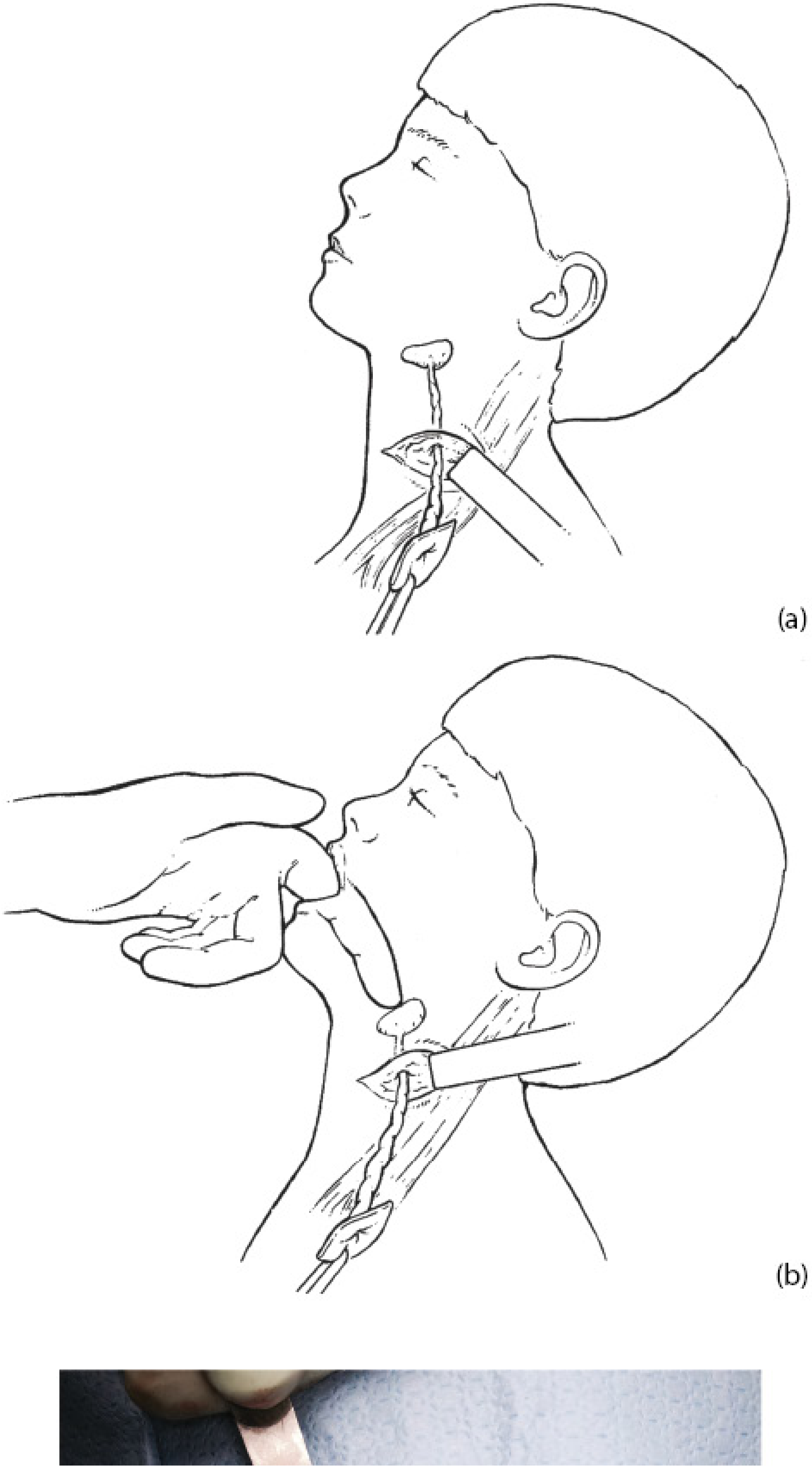
The thyroid gland is exposed through a standard collar incision, and the left lobe is mobilized. The recurrent and superior laryngeal nerves and parathyroid glands should be identified and protected (**Figure 2.9**). If no discrete cyst or tract is found, the fistula may be located at the laryngeal level near the cricothyroid membrane. The fibers of the inferior constrictor muscle are bluntly spread to expose the piriform recess. Extreme caution should be exercised in this region to preserve the external branch of the superior laryngeal nerve. The tract usually passes inferiorly, external to the recurrent laryngeal nerve along the trachea to the superior pole of the thyroid. It may end blindly near the gland or actually penetrate the capsule to terminate in the parenchyma of the left thyroid lobe. Thyroid lobectomy or resection of the superior pole is carried out as indicated by the extent of the cyst.

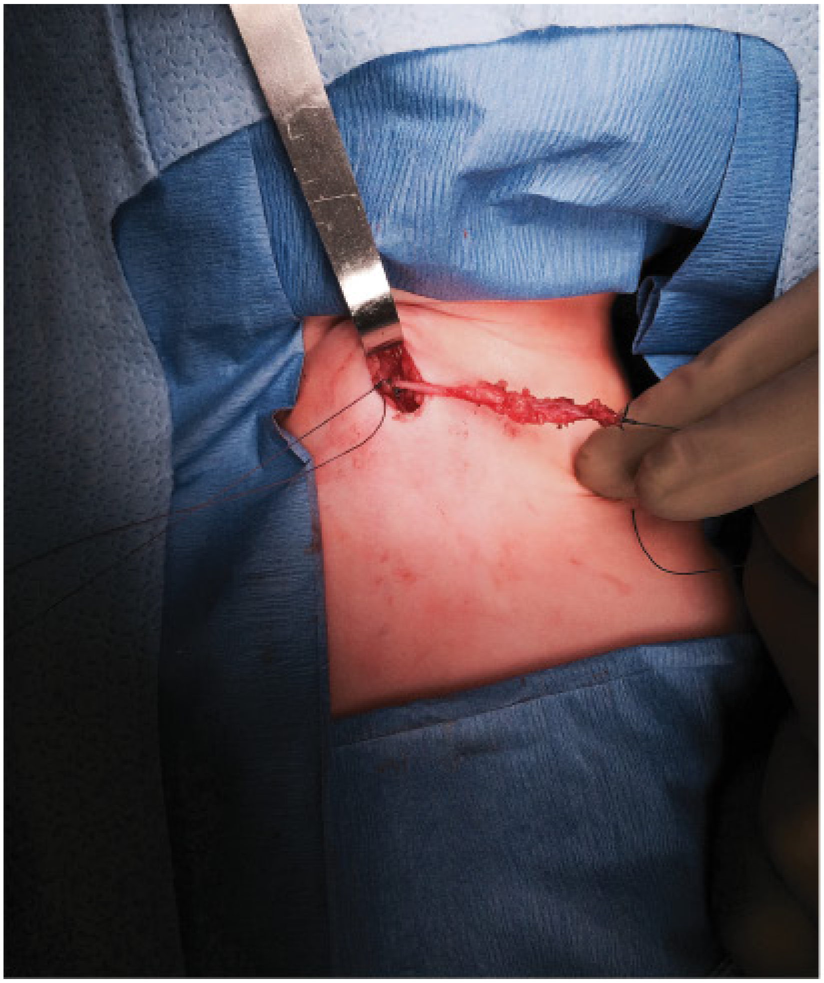

Pyriform sinus
Sinus tract
Superior laryngeal n.
Cyst compressing thyroid gland
Recurrent laryngeal n.

2.9

Excision of a second branchial cleft sinus

The operation to excise a second branchial cleft sinus begins with an elliptical transverse incision at the sinus opening (**Figure 2.10a**), and cephalad dissection of the tract to its furthest extent, generally at the level of the tonsillar pillar (**Figure 2.10b**). The dissection is kept directly on the tract to avoid injury to contiguous structures, such as the internal jugular vein, the bifurcation of the carotid artery, and the hypoglossal nerve. The operation can often be carried out through a single elliptical incision if the tract is kept under gentle traction (**Figure 2.10c**). However, a stepladder incision may be required as the tract is dissected superiorly. Dissection of the sinus tract is facilitated by passing a fine lacrimal probe or injecting saline into the tract via a 24-gauge angiocath.


(a)
(b)


(c)

2.10a–c

Minor branchial remnants

A branchial arch remnant may also occur along the lower anterior border of the SCM near the sternoclavicular joint, and typically consists of a small cartilaginous mass presenting in the subcutaneous tissue. The lesion is usually visible and palpable, and bilateral occurrences are common. An accompanying sinus or cyst is seldom present, and infection is uncommon. Excision may be carried out for cosmetic reasons or may be delayed indefinitely.

POSTOPERATIVE CARE

The operations described are carried out as outpatient procedures unless unusual difficulties are encountered. Antibiotics are not necessary postoperatively. The wound is kept dry for 48 hours. If a drain is placed, it is removed whenever drainage has ceased.

COMPLICATIONS

- *Infection*: 5–10%
- *Recurrence*: 2–12%
- *Facial nerve injury (first branchial)*: 10–40%

OUTCOME

The outcome for the procedures described is usually excellent, both functionally and cosmetically. Operative injury to related anatomical structures is rare and should not occur provided the surgeon has adequate knowledge of the anatomy and that meticulous hemostasis is obtained during dissection to ensure a clear field. Failure to excise the cyst or sinus completely may lead to its recurrence, in which case the patient should be treated with antibiotics and a thorough diagnostic re-evaluation should be initiated.

THYROGLOSSAL DUCT CYSTS

HISTORY

The thyroglossal duct was first described in 1723 by Vater, who called it the "lingual duct." It was later referred to as the "canal of His" following his descriptions in 1855 and 1891. Walter Ellis Sistrunk (1880–1933, American surgeon at the Mayo Clinic, MN) described the basis for the current operation, removing not only the cyst but also the antecedent pathway of thyroid descent to the back of tongue and middle of the hyoid bone.

PRINCIPLES AND JUSTIFICATION

Development of the thyroid gland and thyroglossal tract

The thyroid gland develops as a median thickening of the floor of the pharynx at the level of the second branchial arch (tuberculum impar), during the fourth week of gestation. It descends to its final position in the neck, leaving the thyroglossal duct extending caudally from the foramen cecum of the tongue to the pyramidal lobe of the thyroid, passing anteriorly through or posterior to the hyoid bone. Early in the fifth week of gestation, the attenuated duct loses its lumen and shortly afterwards breaks into fragments. Thyroid remnants may be found along the course of the thyroglossal duct.

Thyroglossal cysts

Thyroglossal cysts occur in about 5% of the population and are the most common anterior cervical swelling in children. They frequently arise just inferior to the level of the hyoid bone. Occasionally, the duct deviates anterosuperiorly once it has passed the hyoid bone, giving rise to a thyroglossal cyst in the submental triangle, where it may be mistaken for a dermoid cyst. Although dermoid cysts may occur below the hyoid bone, they are much more common in the submental triangle and can be distinguished from thyroglossal cysts by their softer, “putty-like” consistency. Occasionally, aberrant thyroid glandular tissue is found along the course of the thyroglossal duct.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

In thyroid hypoplasia, a small central area of aberrant ectopic thyroid tissue may be mistaken for a thyroglossal cyst, and it is recommended that the precise location of the thyroid gland be determined preoperatively. If the thyroid is palpable, no further studies are needed. If the clinician is unable to clearly palpate the thyroid, an ultrasound of the gland should be obtained. Isotope scanning is rarely necessary. Formal thyroid biochemistry is not required unless the child shows signs and symptoms of hypothyroidism. In that case, a thyroid stimulating hormone (TSH) level should be sufficient.

Antibiotics are not necessary if there was no preoperative infection. If the child had a previous abscess at the site of the cyst, a first-generation cephalosporin may be used.

OPERATION

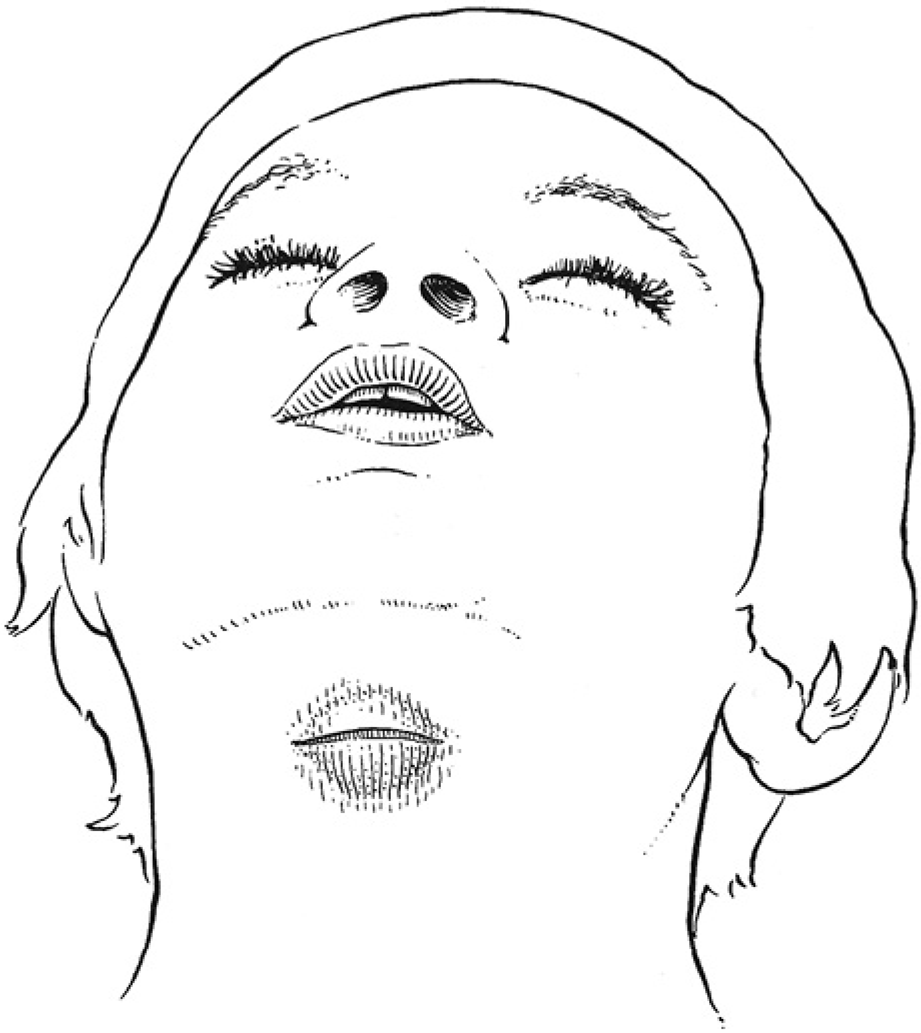
The aim of surgery is to remove the entire thyroglossal duct, including the central part of the body of the hyoid bone, to the level of the foramen cecum. Complete excision is essential to prevent recurrence and eliminate the risk of malignant degeneration. All thyroglossal cysts should be excised to avoid the risk of infection, which makes subsequent surgery more difficult and increases the morbidity and recurrence rates. The operation may be performed on an outpatient basis provided meticulous hemostasis has been achieved.

Position of patient

The child is placed supine with the head extended and the shoulders elevated on a gel roll. The arms are tucked and appropriately padded. A cervical local anesthetic field block with 0.25% bupivacaine diminishes postoperative pain. A chlorhexidine alcohol skin preparation should be used as it appears to have a lower rate of surgical site infections when compared to povidone iodine.

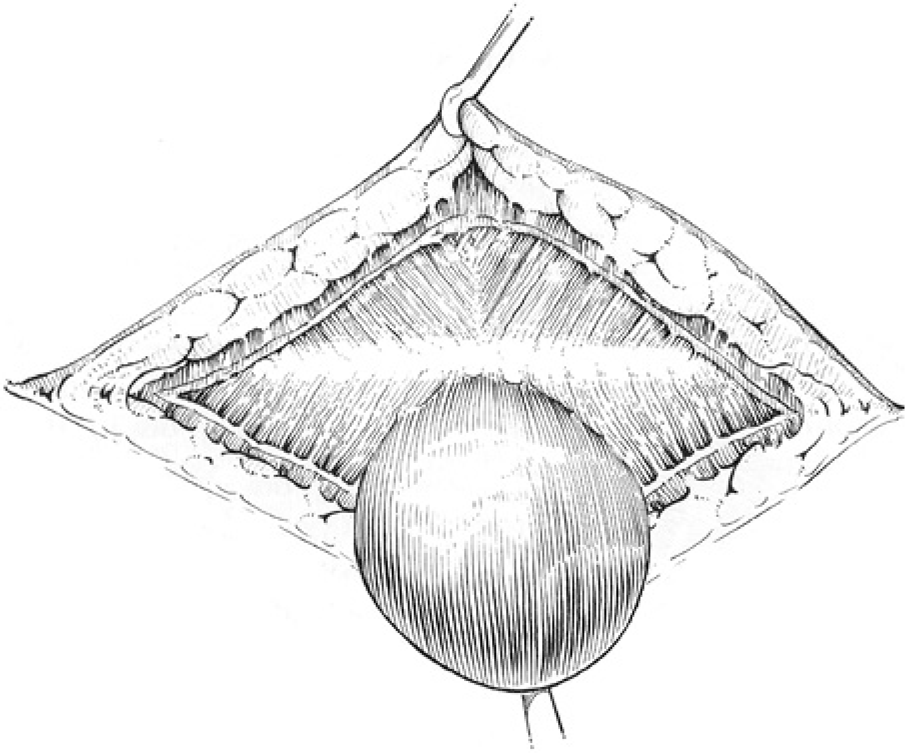
Incision

A short (usually <3 cm) transverse skin crease incision is made over the main prominence of the cyst (**Figure 2.11**).



2.11

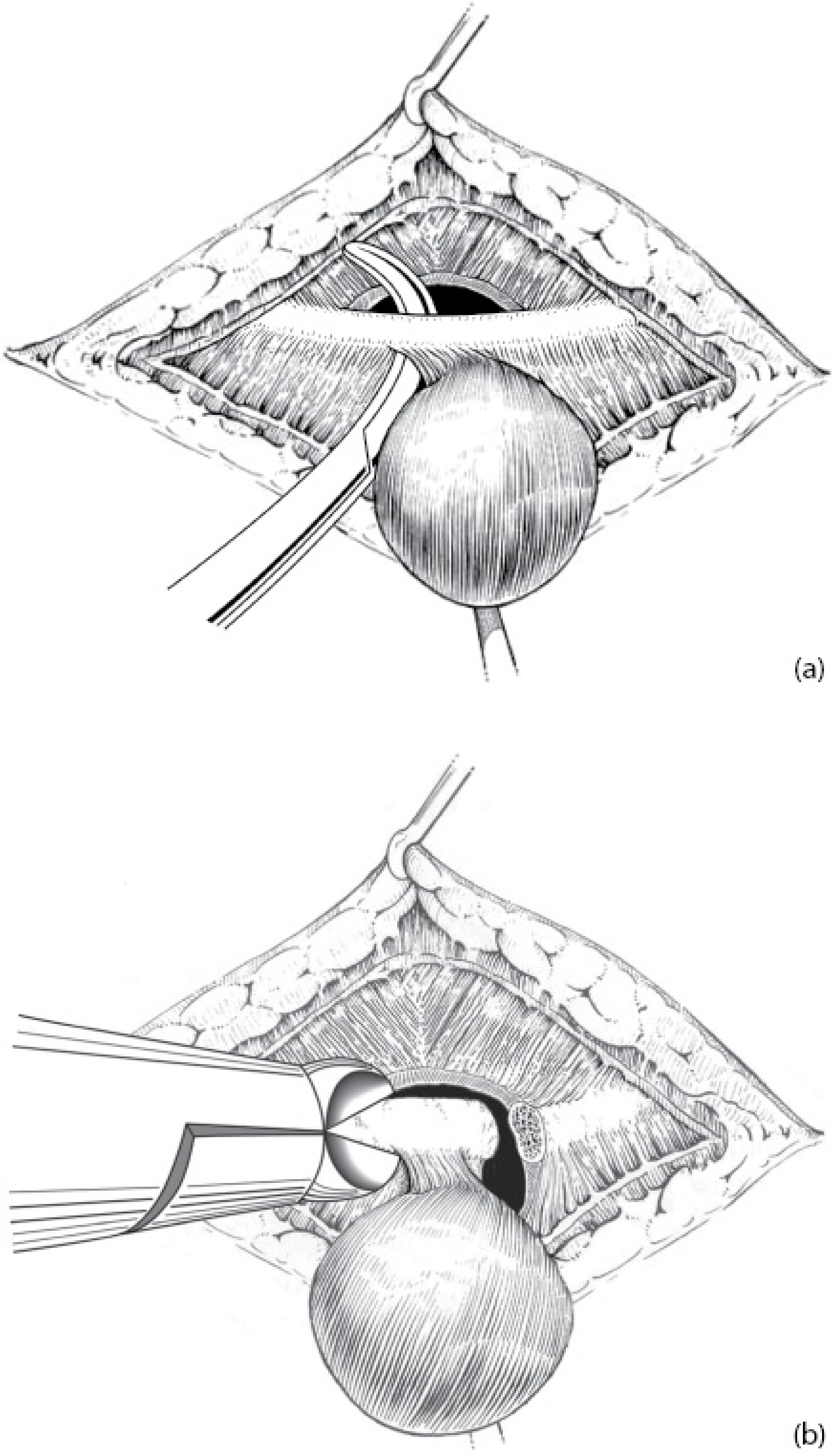
Subcutaneous fat, platysma, and deep cervical fascia are incised in the line of the incision with a diathermy needle. Subplatysmal flaps are raised and a spring-loaded self-retaining retractor is placed. The cyst is freed from its superficial attachments by a combination of sharp and blunt dissection (**Figure 2.12**).



2.12

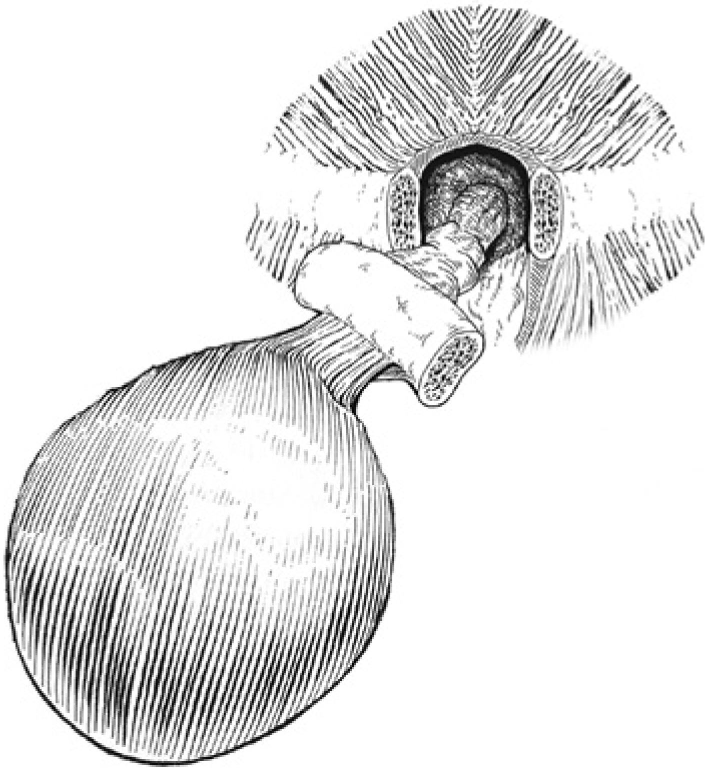
Dissection

The thyroglossal tract is identified at its deep attachment to the cyst and followed between the sternohyoid muscles to the hyoid bone. The centrum of the hyoid bone is freed from the sternohyoid muscles below and the mylohyoid and geniohyoid muscles above with a diathermy needle (**Figure 2.13a**). Small bone-cutting forceps or strong Mayo scissors are then used to divide the body of the hyoid 5 mm to either side of the midline (**Figure 2.13b**). This maneuver is facilitated by grasping and steadying the bone with Kocher clamps.



2.13a,b

A cylinder of geniohyoid and genioglossus muscle 0.5 cm wide, including the duct, is excised back toward the foramen cecum (**Figure 2.14**) and is best performed using needle diathermy. It has been suggested that the dissection is made easier if the anesthesiologist uses a finger to depress the foramen cecum into the wound, but this is seldom of practical value and is a potential danger to the anesthesiologist. The tract is then suture ligated with an absorbable suture.



2.14

Wound closure

The strap muscles are approximated in the midline using absorbable sutures (e.g. 3/0 polyglycolic acid). It is not necessary to reconstitute the hyoid bone because its cut ends tend to be approximated by the muscle sutures. If adequate hemostasis has been secured, no drains are required.

OUTCOME

Recurrence rates of 10–20% have been reported in patients treated by cyst excision only. This decreases to <5% if the thyroglossal duct and hyoid bone have been excised.

ACKNOWLEDGMENTS

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FURTHER READING

Preauricular sinus

Bruijnzeel H, van den Aardweg MT, Grolman W, et al. A systematic review on the surgical outcome of preauricular sinus excision techniques. *Laryngoscope* 2016; 126: 1535–44.

Currie AR, King WW, Vlantis AC, et al. Pitfalls in the management of preauricular sinuses. *Br J Surg* 1996; 83: 1722–4.

Scheinfeld NS, Silverberg NB, Weinberg JM, Nozad V. The preauricular sinus: A review of its clinical presentation, treatment, and associations. *Pediatr Dermatol* 2004; 21: 191–6.

External angular dermoid cyst

Hachach-Haram N, Benyon S, Shanmugarajah K, et al. Back to basics: A case series of angular dermoid cyst excision. *J Plast Reconstr Aesthet Surg* 2013; 66: 57–60.

Lopez M, Vermersch S, Varlet F. Endoscopic excision of forehead and eyebrow benign tumors in children. *J Laparoendosc Adv Surg Tech A* 2016; 26: 226–30.

Branchial arch derivatives

LaRiviere CA, Waldhausen JH. Congenital cervical cysts, sinuses, and fistulae in pediatric surgery. *Surg Clin North Am* 2012; 92: 583–97.

Nicoucar K, Giger R, Jaecklin T, et al. Management of congenital third branchial arch anomalies: A systematic review. *Otolaryngol Head Neck Surg* 2010; 142: 21–8.

Nicoucar K, Giger R, Pope HG Jr, et al. Management of congenital fourth branchial arch anomalies: A review and analysis of published cases. *J Pediatr Surg* 2009; 44: 1432–9.

Schroeder JW Jr, Mohyuddin N, Maddalozzo J. Branchial anomalies in the pediatric population. *Otolaryngol Head Neck Surg* 2007; 137: 289–95.

Thyroglossal duct cysts

Brown BM, Judd ES. Thyroglossal duct cysts and sinuses. *Am J Surg* 1961; 102: 494–501.

Coelho A, Sousa C, Marinho AS, et al. Five-years' experience with outpatient thyroglossal duct cyst surgery. *Int J Pediatr Otorhinolaryngol* 2017; 96: 65–7.

Gallagher TQ, Hartnick CJ. Thyroglossal duct cist excision. *Adv Otorhinolaryngol* 2012; 73: 66–9.

Gupta P, Maddalozzo J. Preoperative sonography in presumed thyroglossal duct cysts. *Arch Otolaryngol* 2001; 127: 200–2.

Ross J, Manteghi A, Rethy K, et al. Thyroglossal duct cyst surgery: A ten-year single institution experience. *Int J Pediatr Otrorhinolaryngol* 2017; 101: 132–6.

Sistrunk WE. Technique of removal of cysts and sinuses of the thyroglossal duct. *Surg Gynecol Obstet* 1928; 46: 109–12.

3

Tracheostomy

KATE STEPHENSON AND MICHELLE WYATT

HISTORY

Tracheostomy is one of the oldest documented surgical procedures and was initially used to relieve upper airway obstruction, principally associated with acute airway infections such as diphtheria and croup. Advances in vaccination and pediatric intensive care practices including endotracheal intubation have reduced this need. The majority of pediatric tracheostomies are now performed for long-term airway or respiratory support.

PRINCIPLES AND JUSTIFICATION

Tracheostomy should be performed as an elective procedure using the conventional dissection technique. Percutaneous dilatational tracheostomy, now widely used in adults, is not suitable for children because the airway is small and often precarious; the trachea is soft, flexible, and very mobile; landmarks can be difficult to palpate; and a displaced tube cannot be rapidly replaced.

Indications

- Airway obstruction
  - Congenital anomalies, e.g. laryngeal web, subglottic stenosis

  - Functional obstruction, e.g. bilateral vocal cord palsy, cricoarytenoid joint fixation
  - Failed extubation with increasing subglottic edema and evidence of mucosal damage
  - Severe external trauma
  - Acute infection, e.g. acute epiglottitis or laryngo-tracheobronchitis, if endotracheal intubation is not possible
  - Tumors, e.g. hemangioma, lymphangioma
  - Tracheomalacia or extrinsic compression of the trachea
- Long-term respiratory support
- Clearance of secretions, e.g. in neurological injury or disease
- As a covering procedure to secure the airway for subsequent upper aerodigestive tract surgery

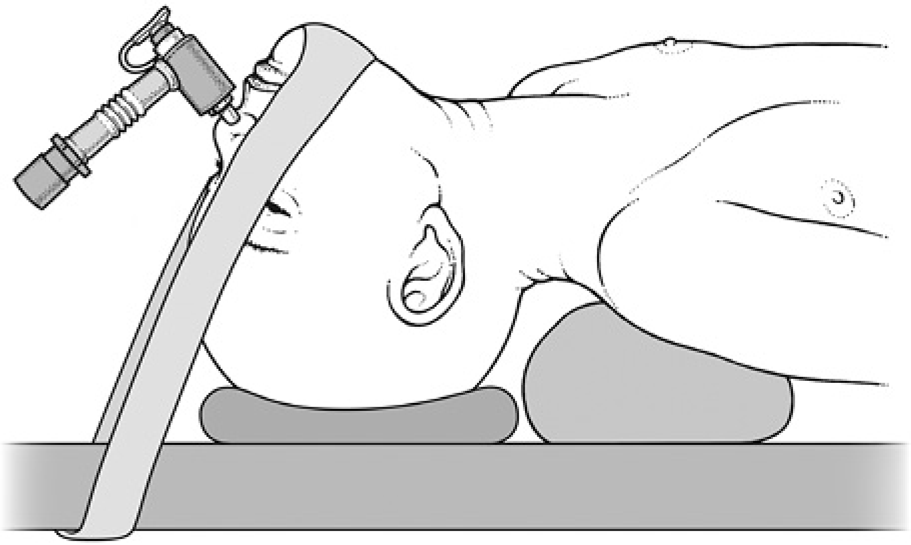
PREOPERATIVE

Anesthesia

The operation is performed under general anesthesia. Children are usually intubated, but in cases where intubation is deemed impossible a laryngeal mask airway (LMA) may be used, or even a facemask. Exceptionally, it might be necessary to undertake the procedure under local anesthesia, using 1% lignocaine (lidocaine) with epinephrine (adrenaline) 1 : 200 000. Tracheostomy in the non-intubated patient is likely to be technically more challenging as the soft pediatric trachea is not reinforced by the endotracheal tube and is less identifiable.

Position of patient

The neck must be extended. A jellyroll or sandbag is placed under the shoulders, and an adhesive tape “chin strap” is applied and attached to the table on either side of the head to stabilize it. The occiput is supported by a head ring. The patient is tipped into a slightly head-up position (**Figure 3.1**).



3.1

Preparation of equipment

Bipolar diathermy and suction should be available. A fenestrated drape is convenient and allows an anesthetic connector to be passed easily beneath it into the surgical field when the tracheostomy tube has been inserted.

A sizing chart, such as that reproduced in **Table 3.1**, should be used to select a tube of the correct diameter and length for the child's age that is best suited to the clinical scenario. The tube is checked and tube-specific suction length measured.

Table 3.1 **Great Ormond Street Hospital chart for pediatric airways**

		Preterm–1 month		1–6 months	6–18 months	18–36 months	3–6 years	6–9 years	9–12 years	12+ years
Cricoid (AP diameter)		3.6–4.8		4.8–5.8	5.8–6.5	6.5–7.4	7.4–8.2	8.2–9.0	10.7	10.7
Trachea (transverse diameter)		**5**		5–6	6–7	7–8	8–9	9–10	10–13	13
Endotracheal tube (Portex®)	**Size = ID**	**2.5**	**3.0**	**3.5**	**4.0**	**4.5**	**5.0**	**6.0**	**7.0**	**8.0**
	OD	3.4	4.2	4.8	5.4	6.2	6.8	8.2	9.6	10.8
Bronchoscope (Storz)	**Size**		**2.5**	**3.0**	**3.5**	**4.0**	**4.5**	**5.0**	**6.0**	
	ID		3.5	4.3	5.0	6.0	6.6	7.1	7.5	
	OD		4.2	5.0	5.7	6.7	7.3	7.6	8.2	
Bivona Uncuffed/Fome-Cuf®/Aire-Cuf®/TTScuff ranges[a]	**Size = ID**	**2.5**	**3.0**	**3.5**	**4.0**	**4.5**	**5.0**	**5.5**		
	OD	4.0	4.7	5.3	6.0	6.7	7.3	8.0		
	Shaft length Neo	30	32	34	36					
	Shaft length Ped	38	39	40	41	42	44	46		
Bivona Hyperflex OD as above	**Size = ID**	**2.5**	**3.0**	**3.5**	**4.0**	**4.5**	**5.0**	**5.5**		
	Max. usable length	55	60	65	70	75	80	85		
Shiley™ *Ped cuffed tube available (PDC)	**Size = ID**		**3.0**	**3.5**	**4.0**	**4.5**	**5.0**	**5.5**	**6.0**	**6.5**
	OD		4.5	5.2	5.9	6.5	7.1	7.7	8.3	9.0
	Length Neo		30	32	34	36				
	Length Ped		39	40	41*	42*	44*	46*		
	Length Ped Long (PDL)						50*	52*	54*	56*

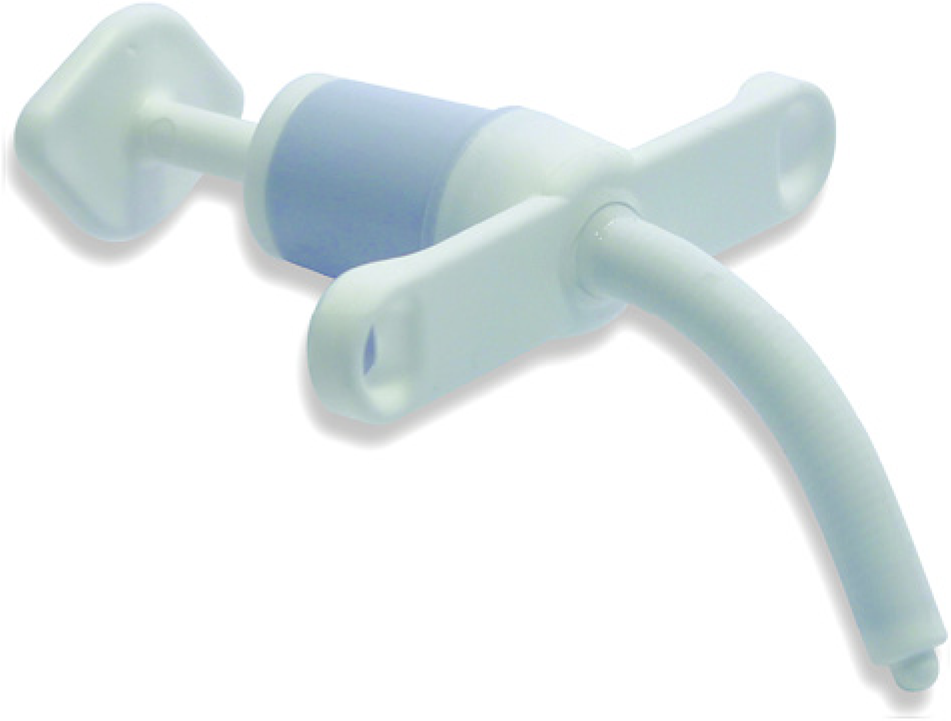
Source: Adapted from Tweedie DJ et al. Paediatric tracheostomy tubes: Recent developments and our current practice. *J Laryngol Otol* 2018; 132(11): 961–8.

Abbreviations: AP, anteroposterior; ID, internal diameter; OD, outer diameter; PDC, pediatric cuffed; PDL, pediatric long; Neo, neonatal; Ped, pediatric.

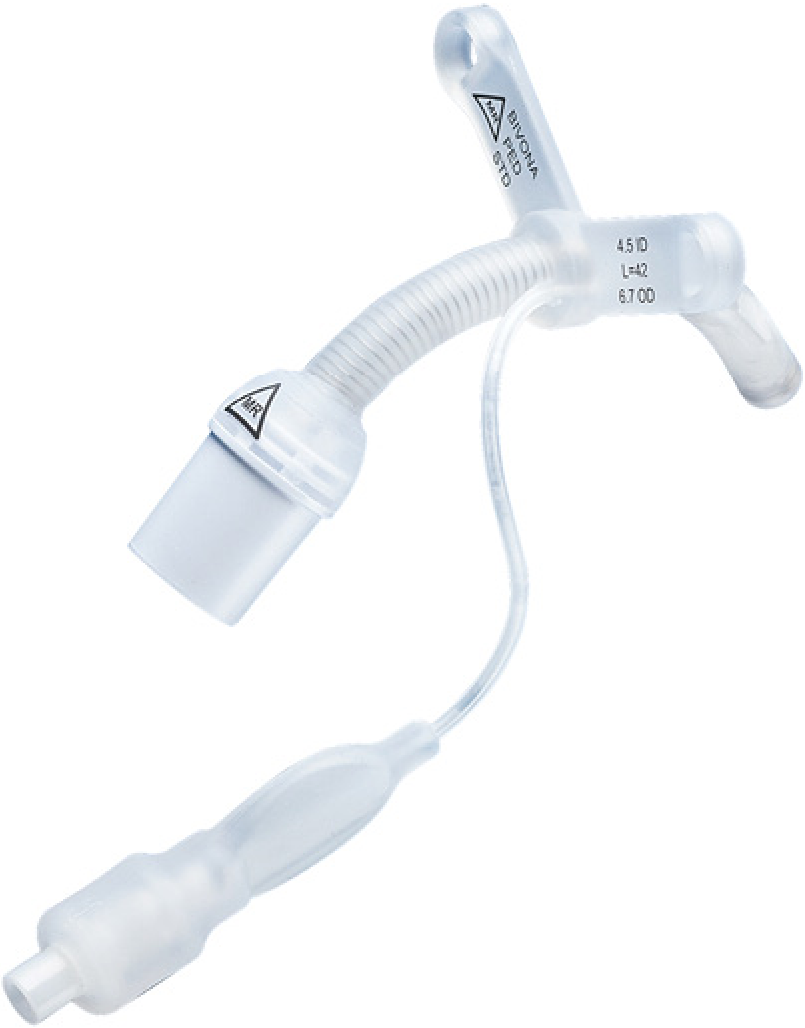
Notes: Suction length should be measured using a suction catheter for each individual tracheostomy tube. All dimensions are in millimeters. Bold highlights tube sizes.

[a]Bivona FlexTend tubes also available in the same sizes.
*Ped cuffed tube available.

Figure 3.2 shows an example of an uncuffed tracheostomy tube (Bivona®). This has a straight neck flange which is well suited to neonates. An example of a tracheostomy tube with a "tight-to-shaft" low-pressure water cuff, a V-shaped neck flange and a flexible extended length proximal shaft (Bivona FlexTend™ TTS™: Smiths Medical, Dublin, OH, USA) is shown in **Figure 3.3**.



3.2

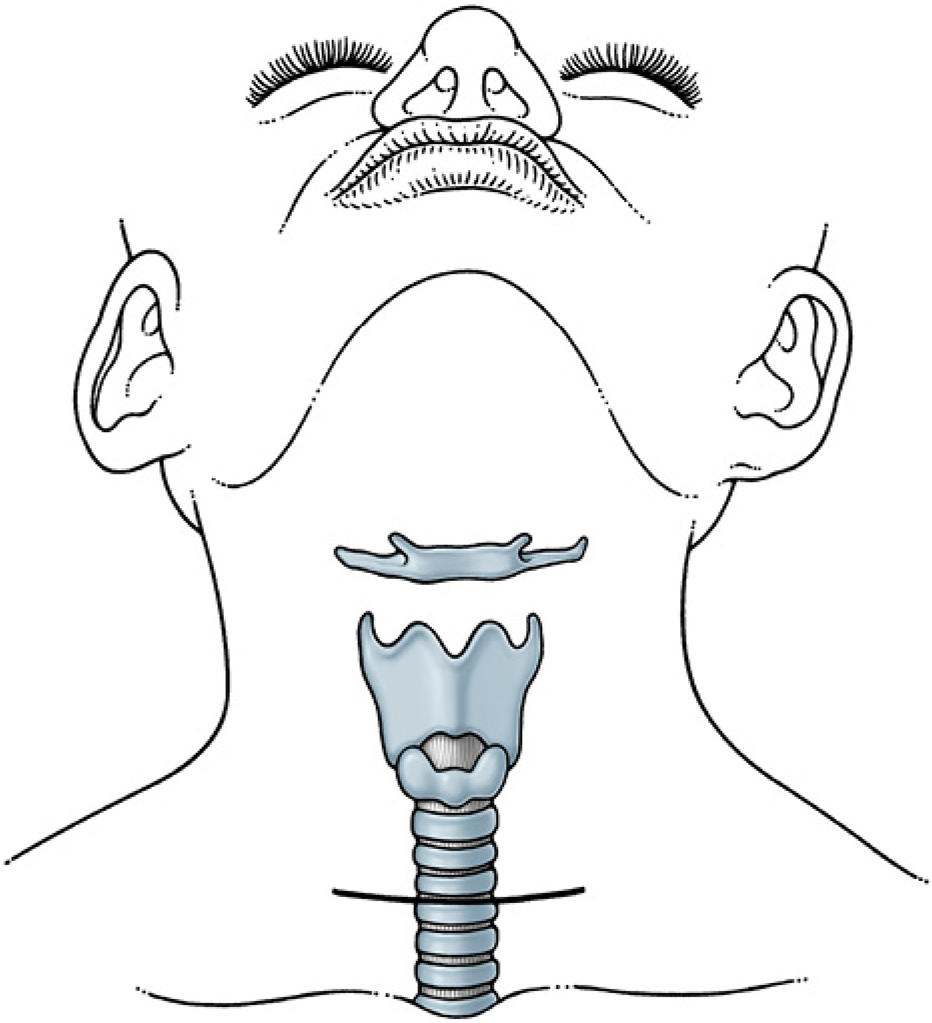


3.3

OPERATION

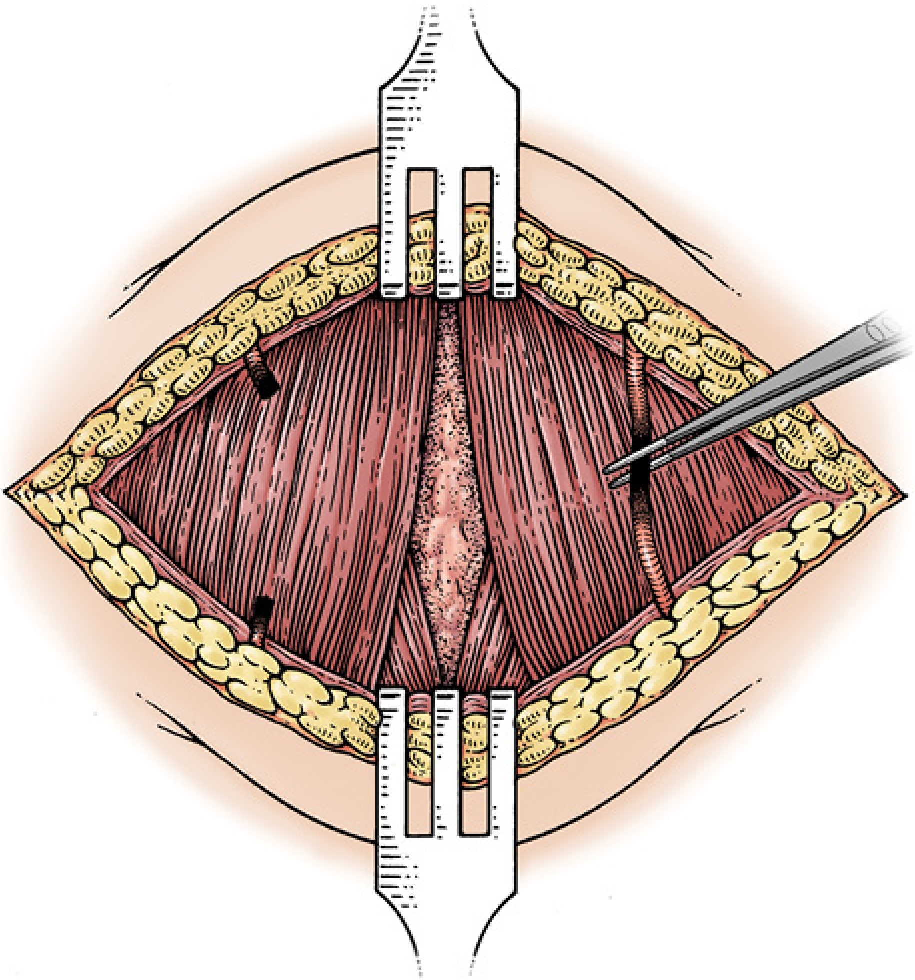
Incision

The neck is palpated carefully so that the hyoid bone, thyroid notch, and cricoid cartilage can be felt and marked. In neonates, the cricoid cartilage is not easily palpable. The skin midway between the cricoid and the suprasternal notch is marked (**Figure 3.4**) and infiltrated using 1% lignocaine with 1 : 200 000 epinephrine. A horizontal skin incision approximately 2 cm in length is made through the infiltrated area, cutting through fat and platysma. Some surgeons favor a vertical incision. In infants and young children, it is advantageous to remove the subcutaneous fat within the incision margins in order to improve access to the trachea; small bipolar diathermy forceps are useful for this purpose.



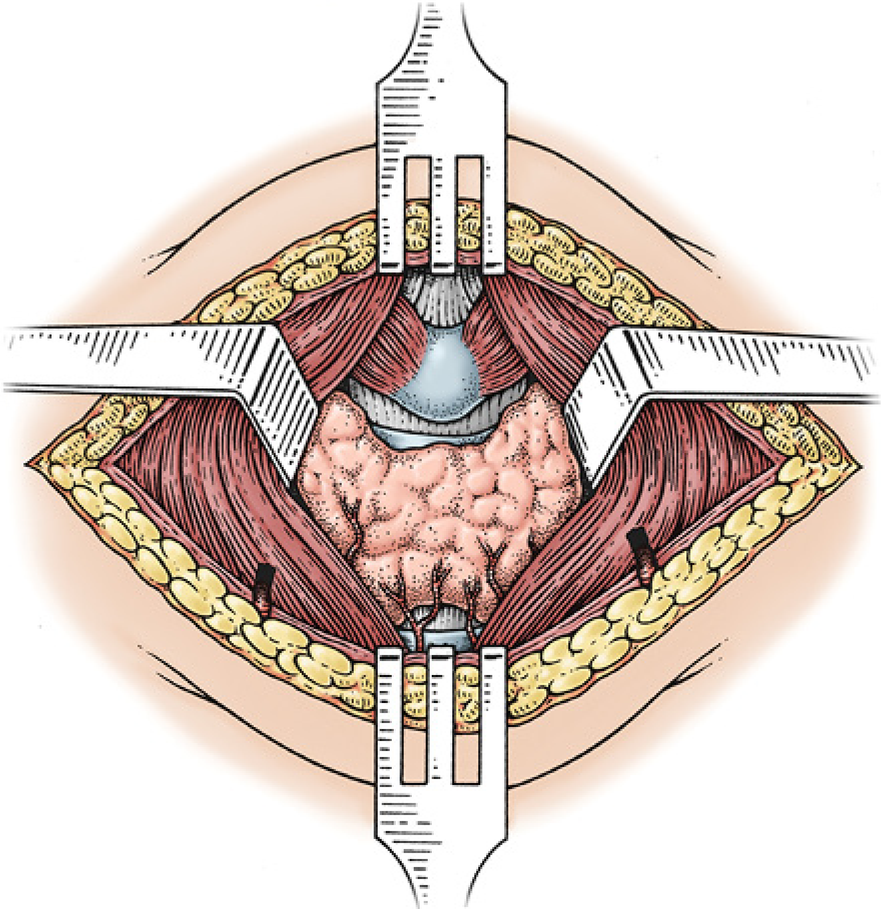
3.4

The incision is deepened in a horizontal plane until the deep cervical fascia investing the sternohyoid and sternothyroid strap muscles is encountered (**Figure 3.5**). Branches of the anterior jugular vein will be seen during this dissection; these should be coagulated and divided with the bipolar diathermy.



3.5

Having identified the strap muscles, a condensation of the investing layer of the deep cervical fascia will be seen running vertically between them in the midline (the cervical linea alba). This plane is entered by blunt dissection and the strap muscles are separated and retracted laterally (**Figure 3.6**). Sprung self-retaining retractors (e.g. Aberdeen) are useful here.



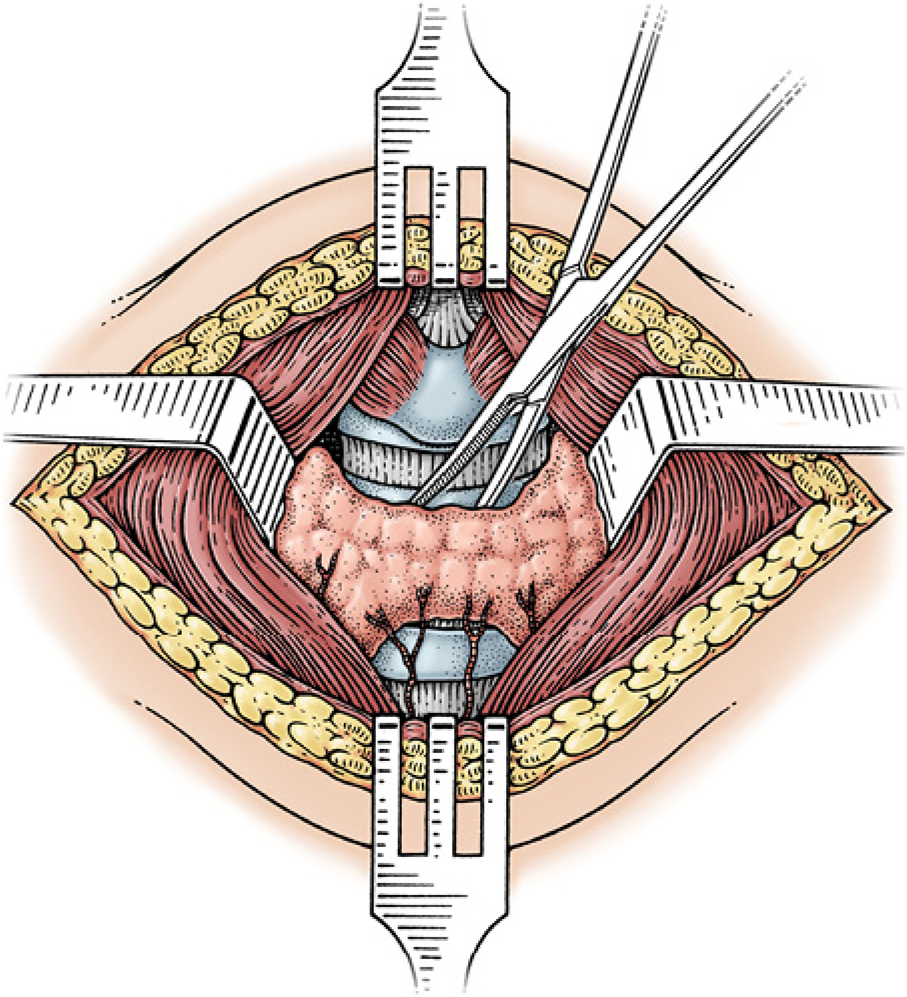
3.6

Exposure of the airway and thyroid isthmus

Retraction of the strap muscles reveals the pre-tracheal fascia and thyroid isthmus joining the two lobes of the thyroid gland. Above the isthmus, the cricoid is seen, although in the neonate it is easier to identify by palpation, feeling for its prominence with fine curved "mosquito" artery forceps. Dissection remains in the midline and particular care is taken in the lower neck where pleura and high brachiocephalic vessels can be encountered.

Dissection of the thyroid isthmus

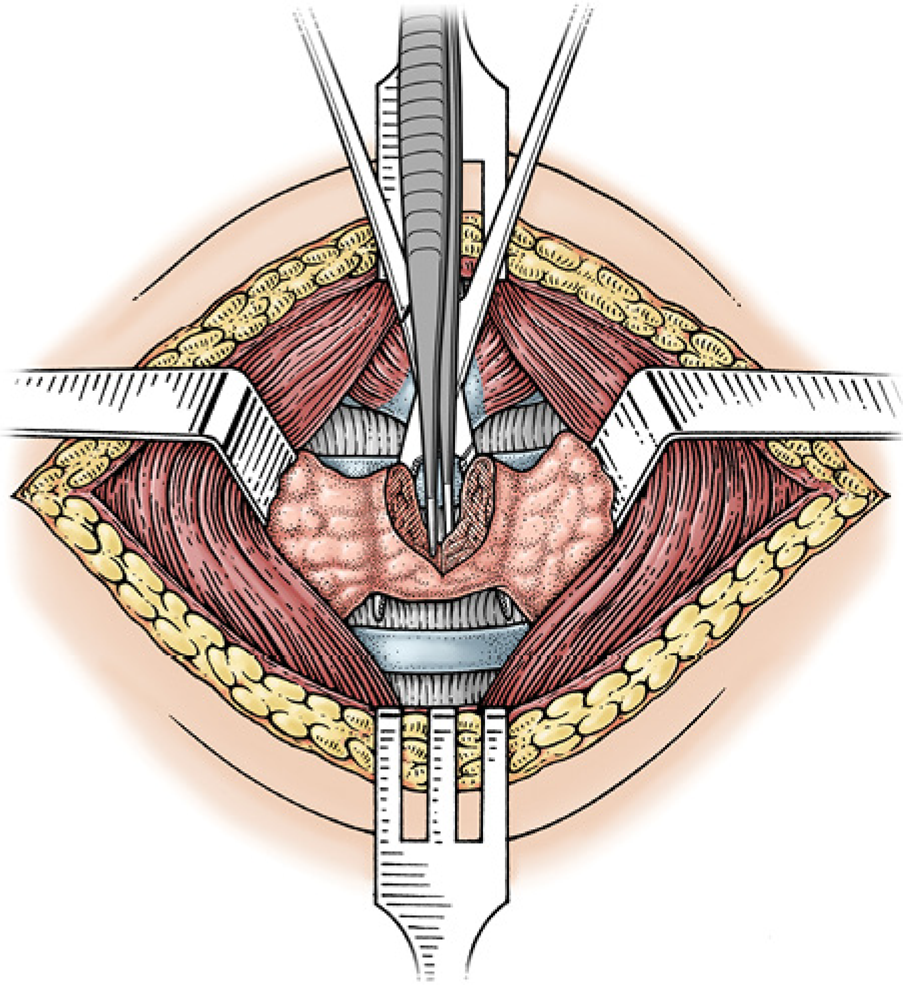
The size and relationship of the thyroid isthmus to the trachea are variable and in almost all cases the isthmus should be divided. A small incision is made through the condensation of the pretracheal fascia at the upper border of the isthmus, and the isthmus is then separated from the underlying trachea by blunt dissection using fine curved artery forceps (**Figure 3.7**). Branches of the inferior thyroid vein will be encountered at the lower border of the isthmus, and these should be coagulated with diathermy.



3.7

Division of the thyroid isthmus

The artery forceps are spread beneath the isthmus, and bipolar diathermy is used to divide it bloodlessly (**Figure 3.8**). In adolescents with a bulky, vascular isthmus it may be preferable to divide the isthmus between hemostats and secure the ends by suture transfixion and ligation.

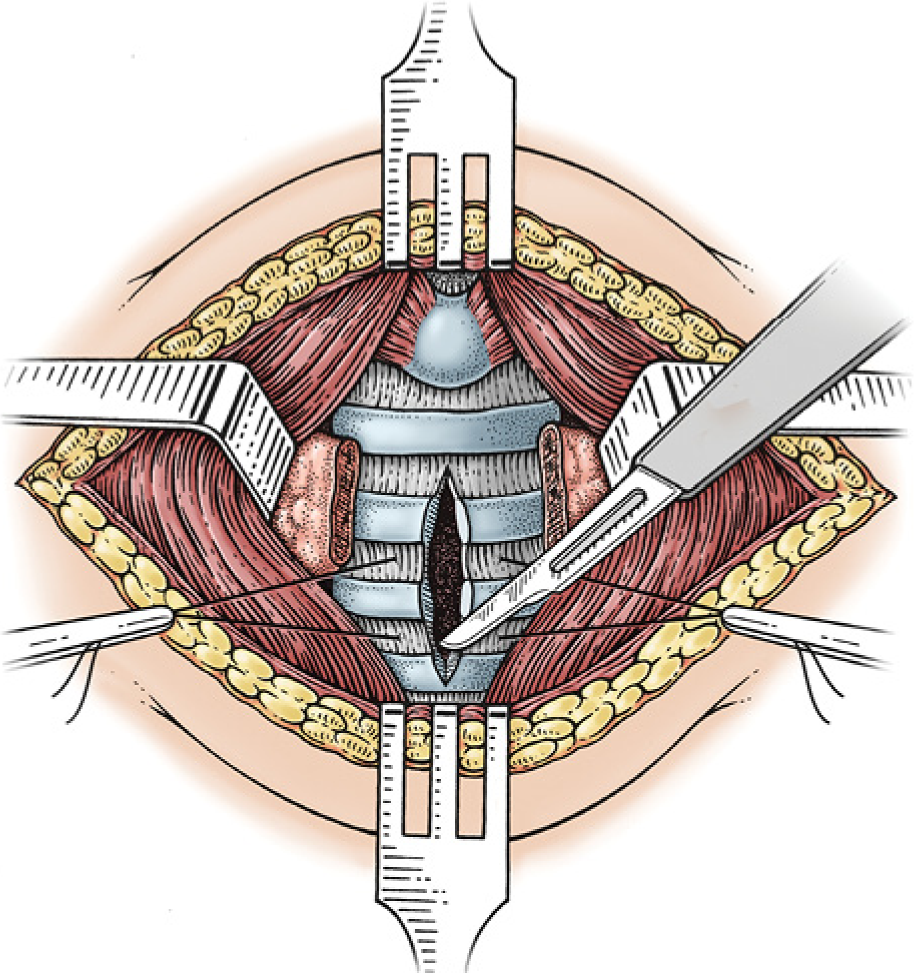


3.8

Opening the trachea

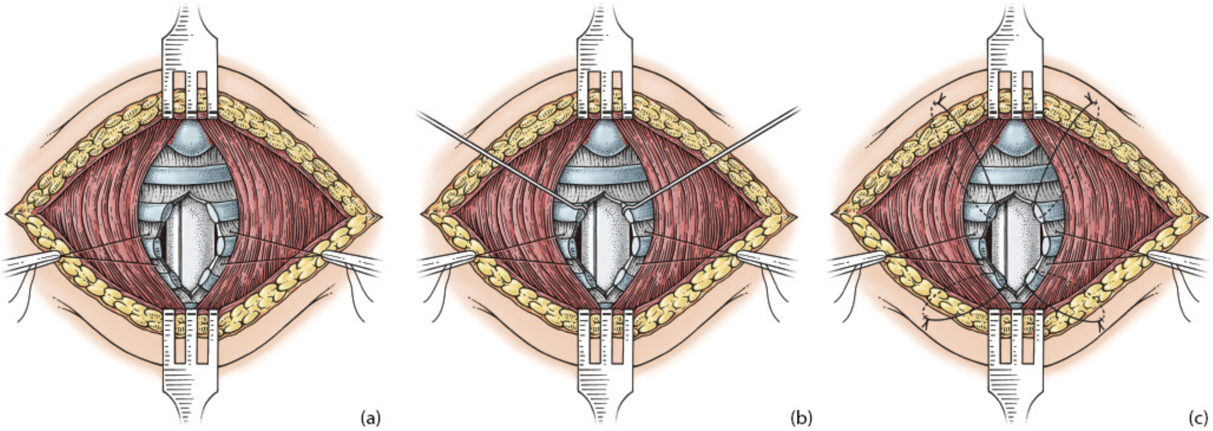
Careful hemostasis is achieved, the cricoid is identified, and the tracheal rings are counted. The first tracheal ring must not be included in the tracheostomy, and in neonates and infants, where distances are small, it is preferable to preserve the second ring as well.

Before making the incision, non-absorbable (e.g. Prolene™) stay sutures are placed to help distract the tracheal opening (**Figure 3.9**). These are later taped to the skin and left in place until the first tracheostomy tube change. For routine tracheostomy in children, a vertical incision is made in the trachea through the third and fourth tracheal rings. If a subsequent reconstructive operation on the larynx is planned, a higher or lower tracheal incision may be considered. Care should be taken with a low tracheostomy that the end of the tracheostomy tube is clear of the carina, and in infants it is important to use a shorter "neonatal"-length tube. A small sucker should be used to prevent blood from entering the trachea.



3.9

The stay sutures are now used to distract the cut edges of the trachea (**Figure 3.10a**), if necessary, aided by fine skin hooks (**Figure 3.10b**). The endotracheal tube will then be clearly seen.

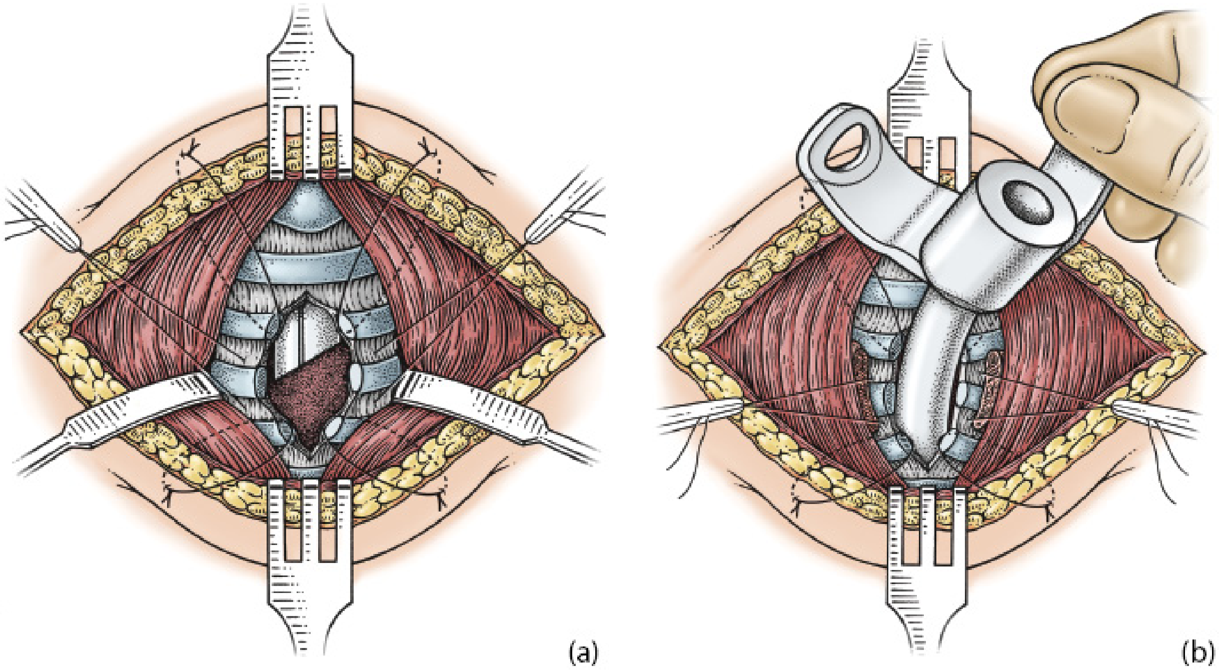


3.10a–c

It is recommended that four absorbable polyglactin (Vicryl™) skin maturation sutures should be used to approximate the skin edges to the margins of the tracheal incision, thus creating a stoma and making it much easier to replace the tube should it become displaced (**Figure 3.10c**).

Insertion of tracheostomy tube

The anesthetist is asked to withdraw the endotracheal tube gently until its tip is just above the tracheotomy (**Figure 3.11a**). The tracheostomy tube is then gently inserted into the tracheal opening in a curved motion, a sterile anesthetic connector is fitted to it, and the end of the connector is passed out of the surgical field beneath the drape to the anesthetist (**Figure 3.11b**).



3.11a,b

Fixation of tracheostomy tube

When the tracheostomy tube is fully inserted, it must be held by the assistant until it is properly secured. If necessary, the skin edges are approximated with a suture on each side of the tube. It is essential to leave a gap around the tube to avoid postoperative surgical emphysema. The stay sutures should be taped to the chest and the tapes clearly labeled “Do not remove” as well as being marked “left” and “right.”

The roll is then removed from beneath the patient’s shoulders and the tracheostomy tube is secured by tying the tapes with a reef knot with the neck flexed (**Figure 3.12**). It is extremely important to remember this neck flexion and to tie the tapes fairly tightly; failure to do this may result in the tube becoming dislodged. Dislodgement of the tracheostomy tube must be avoided during the immediate postoperative period, as its subsequent reinsertion can be difficult in the first few postoperative days. A non-adherent dressing is applied to the wound, with a keyhole to accommodate the tracheostomy tube. This dressing should be kept clean and changed when necessary. At the end of the procedure, the surgeon should listen to the chest with a stethoscope to ensure that air entry is symmetrical; if it is reduced on the left, this suggests that the tracheostomy tube is too long and has intubated the right main bronchus. If necessary, a flexible fiberoptic laryngoscope or bronchoscope may be passed through the tracheostomy tube in order to confirm the position of the tube tip and establish that the tube is of the correct length.

3.12

Technical variations

Bronchoscopy-assisted tracheostomy is an alternative technique. This incorporates an upper airway endoscopy into the procedure and aims to accurately place the tracheostomy below the first tracheal ring while guiding midline dissection.

The *starplasty technique* is based on the principle of a Z-plasty and incorporates skin flaps into a cruciate tracheal incision. This aims to create a stable stoma and reduce the risk of accidental decannulation. It may be particularly suited to patients expected to have a long-term tracheostomy but is associated with the drawback of a high rate of tracheocutaneous fistula.

In rare instances, such as in children with syndromic craniosynostosis, the cartilaginous trachea can be abnormal and the tracheotomy technique altered.

POSTOPERATIVE CARE

The following must be at the bedside:

- A spare tracheostomy tube of the same type and size as that in the patient, and a tube one size smaller (for use if difficulty is encountered replacing the tube with one of the same size); also scissors, spare tapes, and aqueous jelly to lubricate a replacement tube
- Suction apparatus and sterile suction catheters with distal and lateral tip ports
- A fully trained nurse
- A supply of humidified air and an Ambu® bag

A bedside sign giving details of the tracheostomy tube, the upper airway, when the tracheostomy was performed, and emergency procedures is recommended (**Figure 3.13**).

This paediatric patient has a

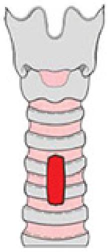
NEW TRACHEOSTOMY

Patient ID: *Patient Label / Details*

Tracheostomy: Add tube specification including cuff or inner tube

______mm ID, ______ mm distal length

Suction: ______ FG Catheter to Depth ______ cm



Indicate on this diagram any sutures in place

UPPER AIRWAY ABNORMALITY: Yes / No

Document laryngoscopy grade and notes on upper airway management or patient specific resuscitation plans

Due 1st tracheostomy change: ___ / ___ / ___ (by ENT ONLY)

In an Emergency: Call 2222 and request the Resuscitation Team and ENT surgeon
Follow the Emergency Paediatric Tracheostomy Management Algorithm on reverse

---

This paediatric patient has a

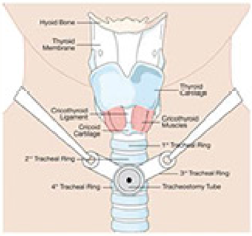
TRACHEOSTOMY

Patient ID: *Patient Label / Details*

Tracheostomy: Add tube specification including cuff or inner tube

______mm ID, ______ mm distal length

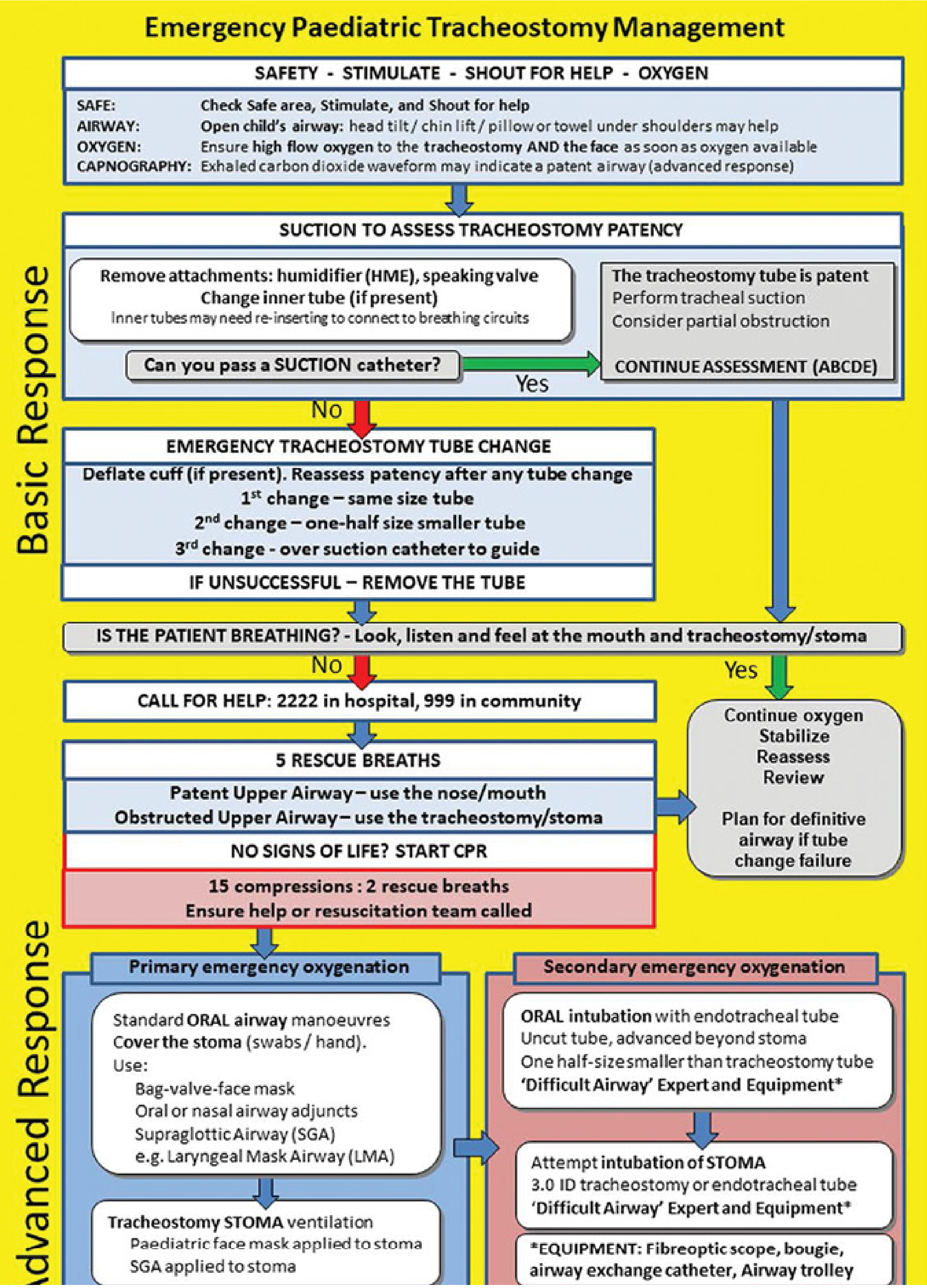
Suction: ______ FG Catheter to Depth ______ cm


Hyoid Bone
Thyroid Membrane
Thyroid Cartilage
Cricothyroid Ligament
Cricothyroid Muscles
Cricoid Cartilage
1st Tracheal Ring
2nd Tracheal Ring
3rd Tracheal Ring
4th Tracheal Ring
Tracheostomy Tube

UPPER AIRWAY ABNORMALITY: Yes / No

Document laryngoscopy grade and notes on upper airway management or patient specific resuscitation plans

In an Emergency: Call 2222 and request the Resuscitation Team and ENT surgeon
Follow the Emergency Paediatric Tracheostomy Management Algorithm on reverse



Emergency Paediatric Tracheostomy Management

Basic Response

SAFETY - STIMULATE - SHOUT FOR HELP - OXYGEN

SAFE:	**Check Safe area, Stimulate, and Shout for help**
AIRWAY:	**Open child's airway:** head tilt / chin lift / pillow or towel under shoulders may help
OXYGEN:	Ensure **high flow oxygen** to the **tracheostomy AND the face** as soon as oxygen available
CAPNOGRAPHY:	Exhaled carbon dioxide waveform may indicate a patent airway (advanced response)

SUCTION TO ASSESS TRACHEOSTOMY PATENCY

Remove attachments: humidifier (HME), speaking valve
Change inner tube (if present)
Inner tubes may need re-inserting to connect to breathing circuits

Can you pass a SUCTION catheter?

Yes

The tracheostomy tube is patent
Perform tracheal suction
Consider partial obstruction

CONTINUE ASSESSMENT (ABCDE)

No

EMERGENCY TRACHEOSTOMY TUBE CHANGE

Deflate cuff (if present). Reassess patency after any tube change
1st change – same size tube
2nd change – one-half size smaller tube
3rd change - over suction catheter to guide

IF UNSUCCESSFUL – REMOVE THE TUBE

IS THE PATIENT BREATHING? - Look, listen and feel at the mouth and tracheostomy/stoma

No

Yes

CALL FOR HELP: 2222 in hospital, 999 in community

5 RESCUE BREATHS

Patent Upper Airway – use the nose/mouth
Obstructed Upper Airway – use the tracheostomy/stoma

NO SIGNS OF LIFE? START CPR

15 compressions : 2 rescue breaths
Ensure help or resuscitation team called

Continue oxygen
Stabilize
Reassess
Review

Plan for definitive airway if tube change failure

dvanced Response

Primary emergency oxygenation

Standard **ORAL airway** manoeuvres
Cover the stoma (swabs / hand).
Use:
- Bag-valve-face mask
- Oral or nasal airway adjuncts
- Supraglottic Airway (SGA)
- e.g. Laryngeal Mask Airway (LMA)

Tracheostomy STOMA ventilation
- Paediatric face mask applied to stoma
- SGA applied to stoma

Secondary emergency oxygenation

ORAL intubation with endotracheal tube
Uncut tube, advanced beyond stoma
One half-size smaller than tracheostomy tube
'Difficult Airway' Expert and Equipment*

Attempt **intubation of STOMA**
3.0 ID tracheostomy or endotracheal tube
'Difficult Airway' Expert and Equipment*

***EQUIPMENT: Fibreoptic scope, bougie, airway exchange catheter, Airway trolley**

NTSP (Paediatric Working Group) www.tracheostomy.org.uk Review January 2020

3.13

A portable chest radiograph should be arranged in the operating room recovery area or immediately on return to the ward, to check the length of the tracheostomy tube with regard to its proximity to the carina, and to ensure that there is no pneumothorax. The tracheostomy tube must be kept clear of secretions by the regular application of suction at hourly intervals for the first 12–24 hours, and thereafter as required. The predetermined suction length is checked and a sterile catheter inserted into the tracheostomy tube gently but swiftly; suction is applied only as the catheter is withdrawn.

Adequate humidification should be provided continuously for the first week, using a mechanical warm-air humidifier for infants and a room-temperature humidifier for older children. This will reduce the tenacity of the secretions and allow them to be cleared by suction more easily. After the first postoperative week, a heat and moisture exchange (HME) device should be used. This consists of multiple layers of water-repellent paper or foam membranes; heat and moisture are trapped during exhalation and then act to warm and humidify inhaled air. There are several types of HME, tailored to the child's age and tidal volume.

The tracheostomy tapes and dressing are changed daily, and the tracheostomy tube is changed routinely after 7 days, at which time the stay sutures are removed. However, if at any time difficulty is encountered during suction, or if breathing becomes noisy or obstructed despite suction, the tube must be changed immediately. This can sometimes be difficult with a new tracheostomy and should be done with adequate help. The child's neck must be extended with a rolled towel under the shoulders, good illumination is essential, and a headlight is desirable. As the tracheostomy tube is withdrawn, the stay sutures are pulled to distract the edges of the tracheal opening and bring them up to the surface of the neck. The tracheostome should thus be clearly visible (especially if the subcutaneous fat was reduced at the time of surgery and skin maturation sutures were placed) and a new tube is inserted under direct vision. If difficulty is encountered, a tracheal dilator is too large to be useful in small children; it is better to insert a suction catheter into the tracheal lumen and use the Seldinger technique to “railroad” the new tube over it into the trachea (or use a smaller tube).

The most common cause of difficulty in suction is displacement of the tube into the pretracheal tissues, which may occur if the tapes are too loose, or if the tube is changed and the new tube is inserted incorrectly.

COMPLICATIONS

Intraoperative

- Bleeding may be encountered from an abnormally high left innominate vein. Careful controlled dissection of the structures in the lower part of the neck should avoid this.
- Damage to the cervical pleura may cause pneumothorax or possibly a pneumomediastinum; dissection lateral to the trachea should be avoided.
- Injury to the larynx, trachea, or esophagus should not occur when an elective tracheostomy is performed.
- Accidental decannulation may occur if the assistant does not hold the tube securely or if the tapes are tied too loosely.

Postoperative

- *Blocking and displacement of the tracheostomy tube.* These complications are preventable by careful nursing and attention to detail when tying the tracheostomy tube tapes.
- *Surgical emphysema.* If this occurs, the wound should be opened fully. It is caused by suturing the wound too tightly around the tracheostomy tube.
- *Hemorrhage.* Reactionary hemorrhage from the wound may occur if hemostasis has been inadequate, a risk minimized by the use of bipolar

diathermy dissection. Erosion of the anterior wall of the trachea can occur if a metal tracheostomy tube of incorrect curvature is used. The overlying innominate artery may also be damaged, in which event a frequently fatal hemorrhage will result.

- *Postoperative pneumonia.* This risk is small if a sterile technique is used for tracheal toilet and effective humidification is maintained. Adequate physiotherapy to encourage coughing is also desirable.
- *Stenosis.* This may occur at the tracheostome if cartilage is removed during the tracheostomy, or at the tip of the tube if suction is too vigorous or incorrectly applied. If the tracheostomy is placed too high and the cricoid cartilage is damaged by the tube, a subglottic stenosis may ensue.
- *Granuloma formation.* A suprastomal granuloma may form in the tracheal lumen at the upper margin of the tracheostome where it is irritated by the tube. If it becomes large enough to interfere with tube changes, phonation, or decannulation, it should be removed either with punch forceps via the stoma or endoscopically. A tube-tip granuloma may arise as a result of irritation of the tracheal wall by either the tip of an ill-fitting tracheostomy tube or unskilled and over-vigorous suction.
- *Suprastomal collapse.* Some degree of suprastomal collapse of the anterior tracheal wall at the upper margin of the tracheostome is almost inevitable in long-term pediatric tracheostomies. Usually it is mild, but occasionally it is sufficiently severe to hinder eventual decannulation. When the suprastomal flap occludes up to 50% of the airway, it can be successfully reduced endoscopically. If more than 50% of the airway is occluded, a formal surgical decannulation may be needed, with anterosuspension of the suprastomal flap or even stomal reconstruction using a cartilage graft.
- *Difficult decannulation.* In the absence of persisting pathology, decannulation is most safely undertaken by progressively downsizing the tube to one of 3.0 mm inner diameter, blocking it for 24 hours, and then removing it if the child can manage comfortably both awake and asleep with the blocked tube *in situ*.
- *Persistent tracheocutaneous fistula.* After prolonged tracheostomy, removal of the tube may result in a persistent fistula at the site of the

tracheostomy in up to 50% of patients. If it fails to close spontaneously and continues to leak mucus, the tract may be excised and a formal closure performed. An upper airway endoscopy is recommended prior to formal closure to ensure that the upper airway is favorable.

OUTCOME

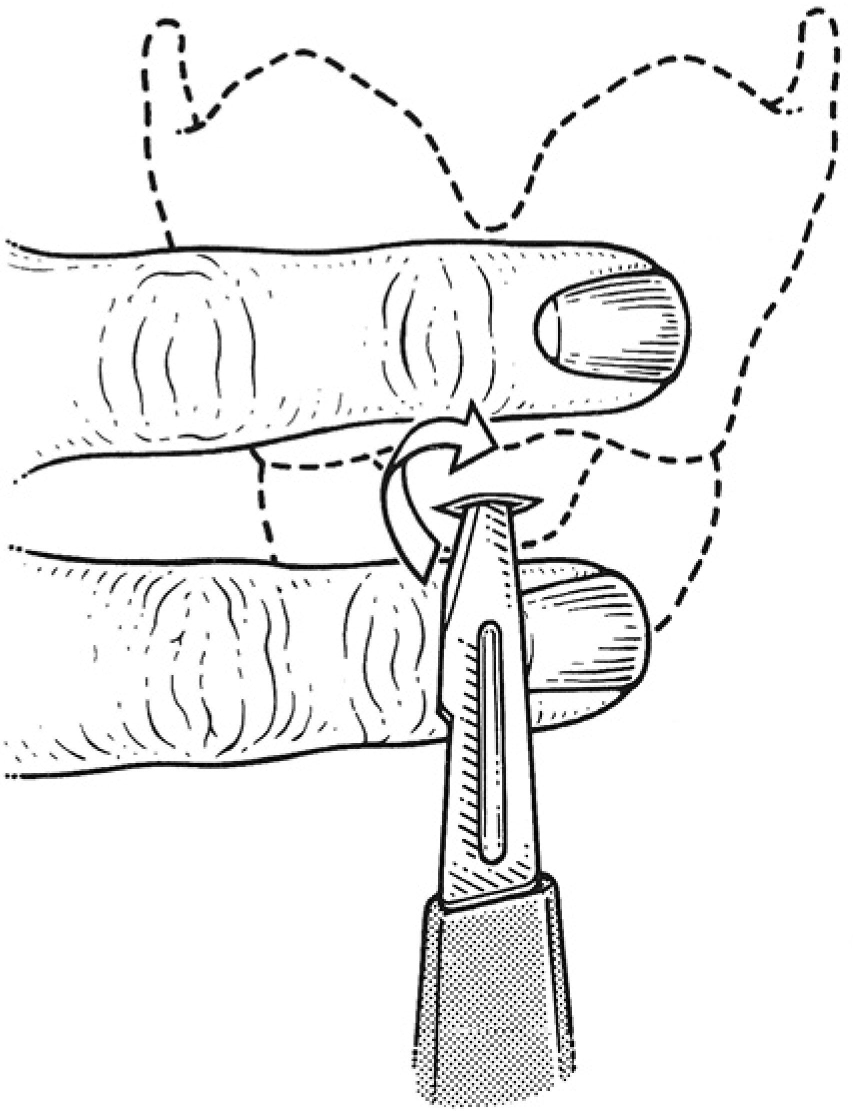
Adverse events associated with pediatric tracheostomy are critical outcomes alongside tracheostomy-related mortality. Results may also be evaluated in terms of length of postoperative hospital stay and decannulation rates; these are likely to be influenced by comorbidities. Coordinated multidisciplinary patient and family-centered care, quality of life, and voice are further important aspects.

CRICOTHYROTOMY

In the rare life-threatening emergency situation when a child cannot be intubated or ventilated, front of neck access is considered. This should ideally be performed by a skilled surgeon. Options include cricothyrotomy or a crash tracheostomy and both can be extremely challenging in the child whose cricothyroid structures and trachea are soft and difficult to identify. Needle and scalpel cricothyrotomy techniques exist, but there is little agreement regarding which is the better technique in the child.

Surgical cricothyrotomy technique

The child is held with the neck extended and the left hand is placed with the first finger over the cricoid cartilage and the second over the thyroid cartilage. A transverse stab incision is made through the cricothyroid membrane (**Figure 3.14**). The blade of the knife is then turned vertically, establishing the airway so that a tube of suitable size may be inserted to preserve the airway and to enable general anesthesia to be undertaken safely. Elective tracheostomy is then performed.



3.14

ACKNOWLEDGMENT

Parts of this chapter appeared in the 7th edition with different authors: C. Martin Bailey and Joe Grainger.

FURTHER READING

Doherty C, Neal R, English C, et al. Multidisciplinary guidelines for the management of pediatric tracheostomy emergencies. *Anaesthesia* 2018; 73: 1400–17.

Koltai PJ. Starplasty: A new technique of pediatric tracheotomy. *Arch Otolaryngol Head Neck Surg* 1998; 124: 1105–11.

Pereira KD, Weinstock YE. Bronchoscopy assisted neonatal tracheostomy (BANT): A new technique. *Int J Pediatr Otorhinolaryngol* 2007; 71: 211–15.

Tweedie DJ, Cooke J, Stephenson KA, et al. Paediatric tracheostomy tubes: Recent developments and our current practice. *J Laryngol Otol* 2018; 132: 961–8.

PART II

Endoscopy

4

Bronchoscopy

MARCUS D. JARBOE AND JAMES D. GEIGER

PRINCIPLES AND JUSTIFICATION

Bronchoscopic examination of the pediatric airway may be performed electively for the diagnosis and/or management of airway disease. While flexible bronchoscopy may be regarded as less invasive, rigid bronchoscopy remains the most versatile method of airway assessment and is where this chapter will focus. Urgent rigid bronchoscopy may be required in the case of foreign body aspiration and therefore a good understanding of bronchoscopic technique is necessary for the pediatric surgeon. Rarely, the ventilating bronchoscope may be useful in securing the difficult airway.

Indications

- Diagnostic
  - Airway assessment, e.g. for stridor
  - Assessment of tracheomalacia
  - Diagnosis of tracheoesophageal fistula: H-type or recurrent
  - Biopsy of tracheal/bronchial lesions
  - Culture of secretions
- Therapeutic
  - Removal of foreign bodies
  - Dilatation of stenoses
  - Laser destruction of lesions

- Management of severe upper airway obstruction
- Management of pulmonary/tracheal hemorrhage

PREOPERATIVE

Investigations

Anteroposterior and lateral chest radiographs should be performed in the assessment of foreign body inhalation. Radio-opaque foreign bodies may be observed directly. In cases where the foreign body is radiolucent, air trapping, collapse, or consolidation may be seen. The air trapping may be accentuated with decubitus films of the chest.

Anesthesia

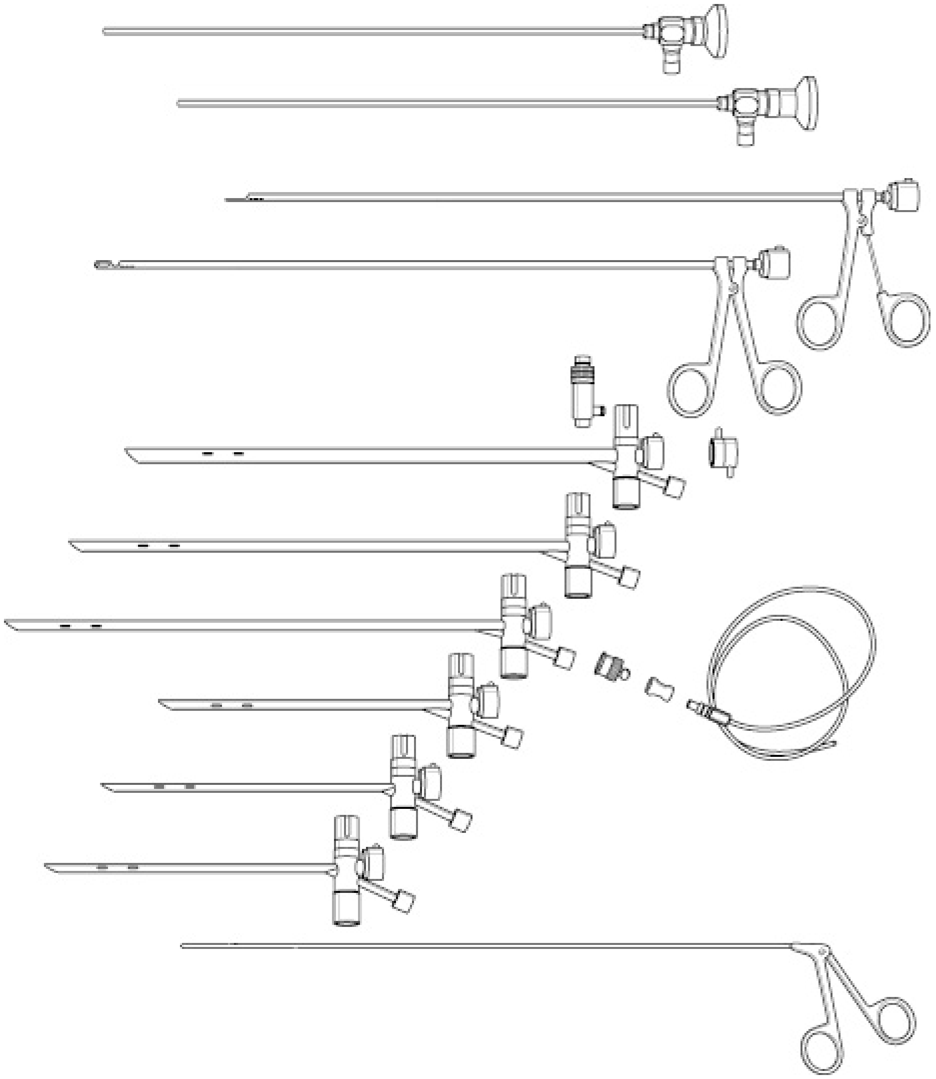
Rigid bronchoscopy is performed under general anesthesia ideally using IV anesthesia with agents such as propofol. Spontaneous ventilation is optimal, enabling a dynamic assessment of airway lesions and is preferred in cases of severe airway obstruction.

Children should be premedicated with atropine in order to reduce airway secretions and facilitate the effective application of topical anesthesia to the larynx and vocal cords. Following the application of topical anesthesia to the larynx to prevent laryngospasm, a nasopharyngeal airway is passed in order to deliver inhalational anesthesia and/or oxygen until the bronchoscope is in place.

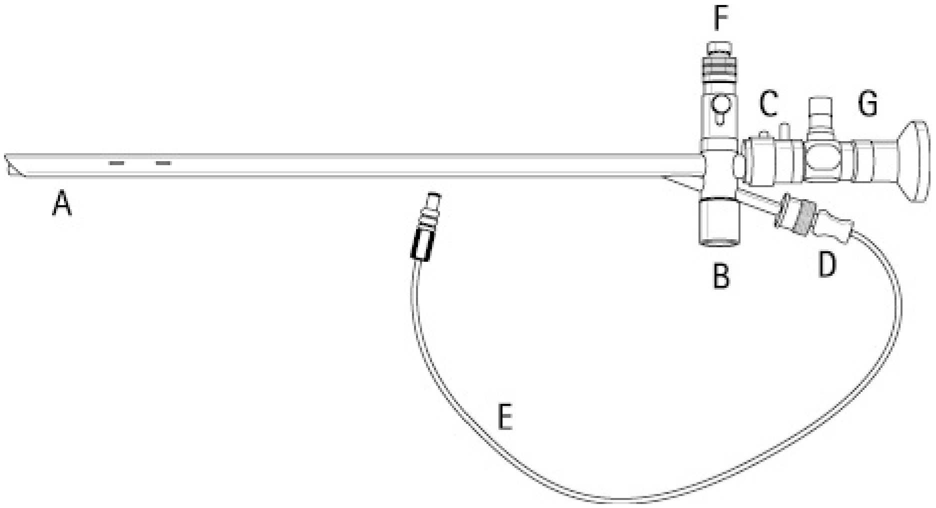
OPERATION

Preparation

The ventilating bronchoscope and any associated ancillary equipment should be prepared prior to the induction of general anesthesia (**Figures 4.1 and 4.2**) (**Table 4.1**). In many instances the airway carts may have mismatching pieces and it is therefore important to have complete sets assembled before induction. When removing foreign bodies, having multiple types of optical forceps available and at the ready is prudent. In patients who are intermediate in size, having multiple sizes of scopes and instruments ready for use can increase efficiency and safety. For diagnostic evaluation in the spontaneously ventilating patient, a rigid Hopkins rod endoscope may be used alone, reducing equipment diameter and therefore reducing mucosal trauma. A defogging solution is used to prevent condensation forming on the lens.



4.1 Storz ventilating bronchoscopes and ancillary parts, Hopkins rod endoscopes, and a selection of grasping forceps.



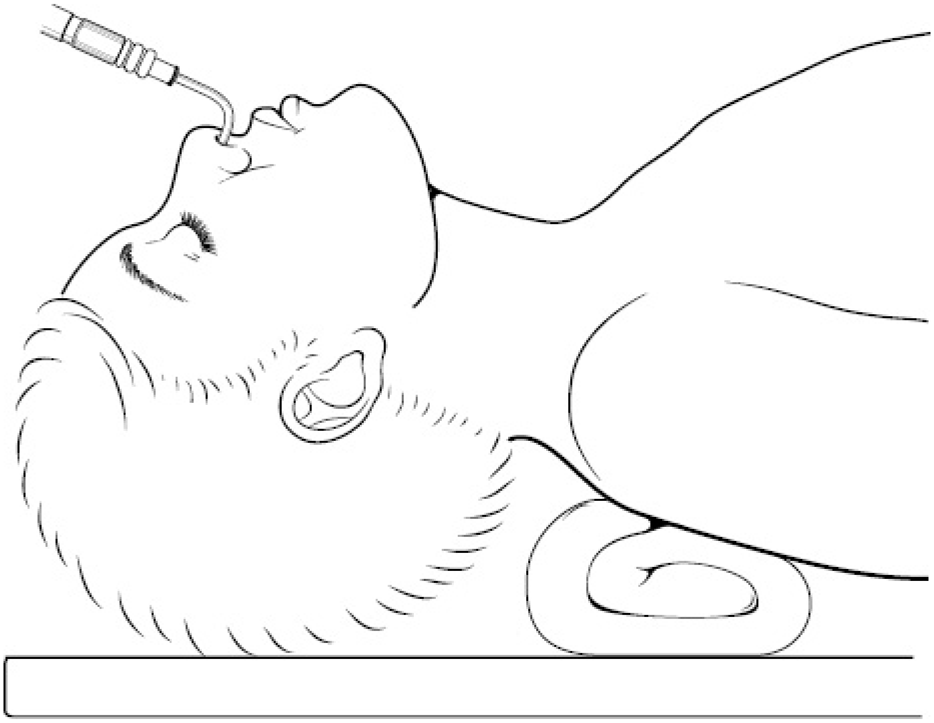
4.2 An assembled Storz ventilating bronchoscope: A, ventilating bronchoscope; B, 15 mm anesthesia port; C, bridge; D, suction post with bung; E, flexible suction tubing; F, prismatic light deflector; G, 0° Hopkins rod endoscope with fiberoptic light port.

Table 4.1 Storz bronchoscope sizes relative to the child's age and airway diameter

		Preterm–1 month	1–6 months	6–18 months	18–36 months	3–6 years	6–9 years	9–12 years	12–14 years
Cricoid (AP diameter)	ID (mm)	3.6–4.8	4.8–5.8	5.8–6.5	6.5–7.4	7.4–8.2	8.2–9.0	9.0–10.7	10.7
Endotracheal tube size	ID (mm)	3.0	3.5	4.0	4.5	5.0	6.0	7.0	8.0
Bronchoscope size (Storz)	Size	2.5	3.0	3.5	4.0	4.5	5.0	6.0	6.0
	ID (mm)	3.5	4.3	5.0	6.0	6.6	7.1	7.5	7.5
	OD (mm)	4.2	5.0	5.7	6.7	7.3	7.8	8.2	8.2

Position of patient

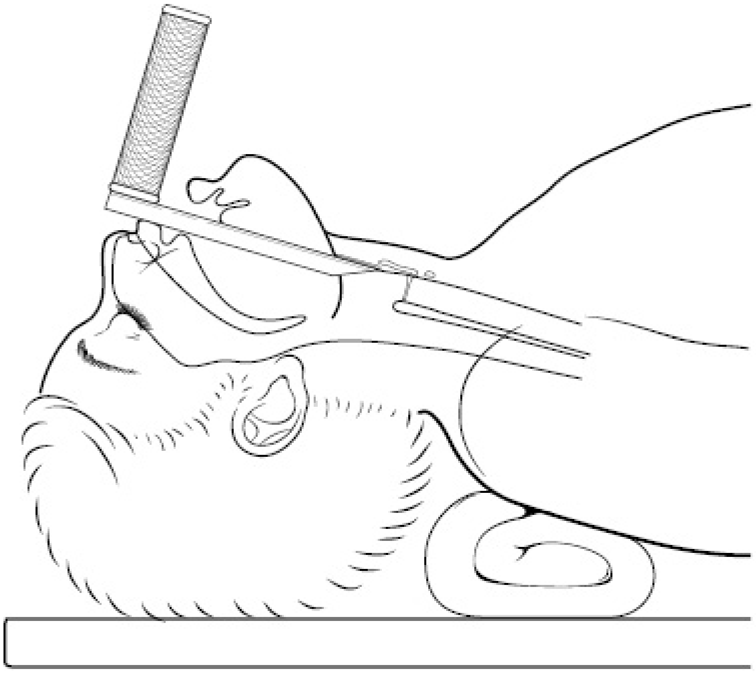
The child should be placed supine on the operating table. The neck should be extended with a shoulder roll and the head supported by a head ring (**Figure 4.3**).



4.3

Direct laryngoscopy

A moistened swab may be used to protect the upper alveolar ridge in infants. In older children, a Silastic gum guard may be preferred. Direct laryngoscopy is performed using an appropriately sized open laryngoscope with a lateral slot. Once the tip of the laryngoscope is in the vallecula, the larynx is exposed by pulling the epiglottis forward (**Figure 4.4**). Using the upper alveolus or dentition as a fulcrum should be avoided.



4.4

Diagnostic bronchoscopy

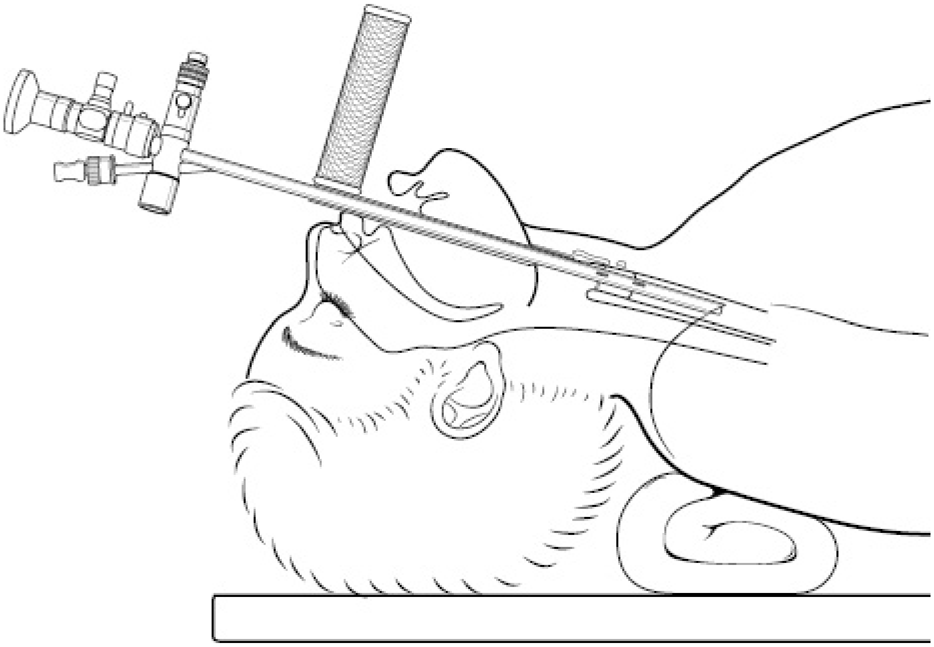
Diagnostic evaluation is performed in a systematic fashion. The bronchoscope is passed distally identifying the carina, which may appear rather broad in the neonate. From the carina, to enter the right main bronchus, the child's head is rotated to the left and simultaneously the bronchoscope is angled to the right. The opposite procedure is followed to enter the left main bronchus.

A fine flexible suction tube may be passed through the suction port of the bronchoscope to clear secretions.

When evaluating for a tracheoesophageal fistula, careful and thorough examination is necessary. Standard type-C fistulas are usually clear but second or higher H-type fistulas can be quite subtle. Gentle probing with a Kumpe catheter +/– an 0.035 angled glidewire can be revealing.

Introducing the bronchoscope

The bronchoscope is inserted through the laryngoscope lumen. It is sometimes helpful to unlock the Hopkins rod from the bronchoscope during insertion and withdraw it slightly into the lumen of the bronchoscope (**Figure 4.5**). As the vocal cords are approached, the bronchoscope is rotated 90 degrees so that its leading edge is in an anteroposterior direction. As the bronchoscope enters the trachea, it is rotated back 90 degrees and the laryngoscope is withdrawn. The fingers of the left hand may be used to support the bronchoscope at the mouth and protect the lips and alveolus. While the bronchoscope is in place, the anesthetic circuit should be connected to the side port of the bronchoscope.



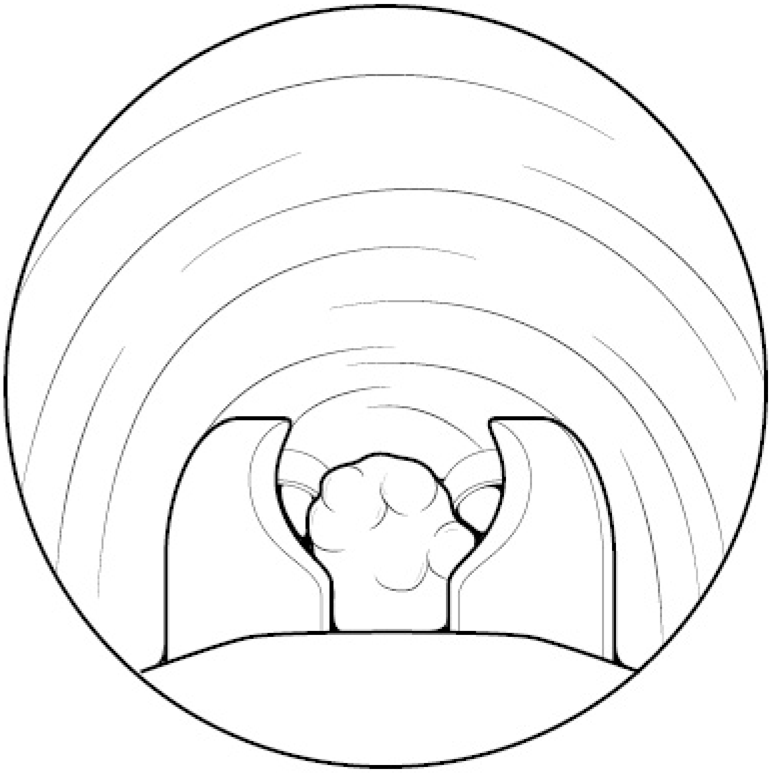
4.5

Foreign body removal

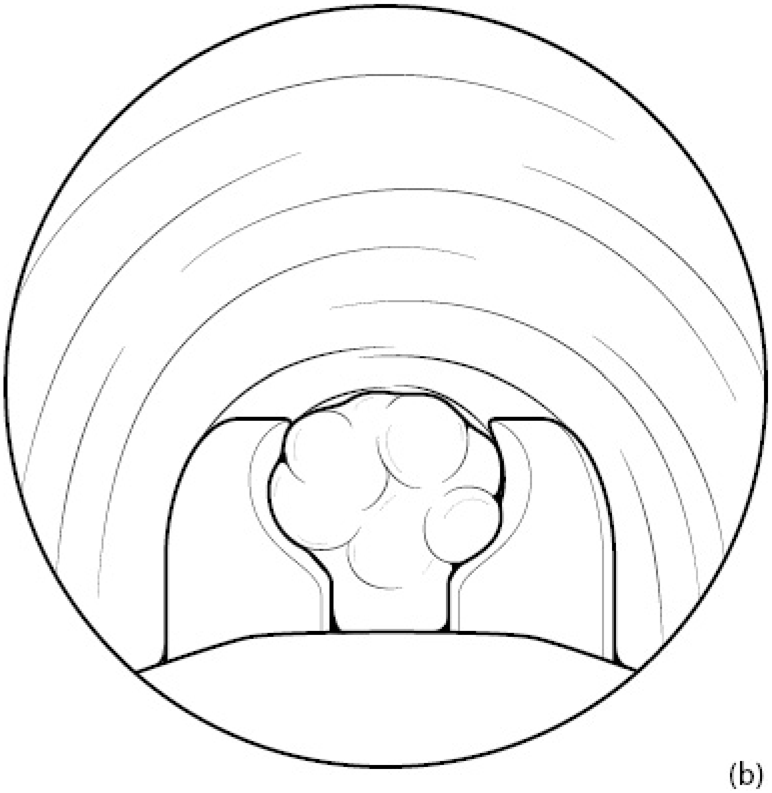
Having identified the foreign body, an assessment is made as to whether the foreign body can be safely removed endoscopically. Dilute epinephrine solution (1–2 mL of 1 : 10 000) may be instilled around the foreign body using the flexible suction catheter, provided there is no contraindication. This reduces mucosal edema and facilitates foreign body removal.

The bronchoscope is left in place and the Hopkins rod endoscope removed from its lumen. An appropriate optical forceps is selected according to the nature of the foreign body. A variety of types of forceps is available, but optical forceps are preferred over flexible side-arm forceps where possible.

The optical forceps are inserted through the bronchoscope, which should be positioned just proximal to the foreign body (**Figure 4.6a**). The foreign body is grasped and drawn into the bronchoscope lumen (**Figure 4.6b**). Larger foreign bodies should be withdrawn to the leading edge of the bronchoscope and then the entire apparatus withdrawn. Alternative approaches include the use of balloon catheters to free up a foreign body or the use of wire stone baskets.



(a)



(b)

4.6a,b

Following removal, a further endoscopic examination should be performed in case of multiple foreign bodies being present.

POSTOPERATIVE CARE

Following recovery from anesthesia, children should be observed. Where bronchoscopy has been performed in the presence of significant airway narrowing, or where there has been significant mucosal trauma, dexamethasone may be administered in the immediate postoperative period to reduce edema and admission may be required.

Respiratory distress following rigid bronchoscopy may be helped by dexamethasone and nebulized epinephrine. However, increasing airway obstruction may necessitate intubation.

After foreign body removal, most children recover rapidly with no long-term sequelae. In cases where a foreign body has been present for some time or where there are radiographic changes suggesting infection, a course of antibiotics may be indicated. In the case of peanut aspiration, a follow-up bronchoscopy might be indicated to rule out the development of a stricture.

COMPLICATIONS

Intraoperative

- Dental injury or damage to the alveolar margin may occur, especially where there is micrognathia. Protection of the teeth and gingivae during the procedure with the operator's fingers and gauze swabs can avoid this.
- Bleeding may be encountered if granulation tissue is removed or mucosal injury occurs. This usually resolves spontaneously, but topical epinephrine may be helpful.
- Airway obstruction can occur while removing a foreign body at the level of the cords. If obstruction occurs, precluding ventilation, use the

bronchoscope to push the foreign body back into the trachea and if necessary, further down into the mainstem bronchus.

- Hypercarbia occurs with prolonged use of the ventilating bronchoscope, especially when smaller sized bronchoscopes (<4.0) are used. Removal of the Hopkins rod endoscope from the lumen every 5 minutes or so can prevent this.

Postoperative

- *Laryngospasm*. This may be minimized by good anesthetic technique, use of topical local anesthetic to the larynx, minimal mucosal contact and ensuring there is no active bleeding at the end of the procedure.
- *Airway obstruction* may occur as a result of trauma to an already compromised airway along with the effects of anesthesia. Nebulized epinephrine and oral or IV dexamethasone may provide time for edema to settle, but in severe cases intubation or tracheostomy may be necessary.
- *Aspiration*. Sufficient time should elapse following the application of topical anesthesia to the larynx before feeding in order to prevent aspiration.

OTHER THERAPEUTIC OPTIONS

A full description of other therapeutic airway techniques is beyond the scope of this chapter. However, the ventilating bronchoscope has many other roles including the management of tracheal stenosis, tracheal and suprastomal granulations, and indwelling stent removal.

In addition to foreign body forceps, balloon catheters and laser fibers may be used down the bronchoscope alongside a Hopkins rod endoscope.

The bronchoscope may also be a valuable adjunct in the management of the difficult pediatric airway where other intubation methods have failed. A far lateral approach to the glottis through the glossotonsillar sulcus can often be successful, provided the operator is familiar with the ventilating bronchoscope.

FLEXIBLE BRONCHOSCOPY

In some cases, particularly where therapeutic intervention is not required and the child's airway not significantly compromised, flexible bronchoscopy may provide a "less invasive" alternative to rigid endoscopy. However, in contrast to adult flexible bronchoscopy, general anesthesia is still required for flexible bronchoscopy in children, reducing some of the benefits seen in adult flexible bronchoscopy. In general, flexible bronchospes can reach further distally in the airway but therapeutic options are limited as compared to the rigid scope.

Flexible bronchoscopy may be used instead of rigid endoscopy for the assessment of tracheobronchial lesions, biopsy of lesions, and bronchoalveolar lavage. Flexible bronchoscopy may be performed in the spontaneously ventilating patient in a similar fashion to rigid endoscopy or through an endotracheal tube or laryngeal mask airway. Performing flexible bronchoscopy through an endotracheal tube ensures a secure airway and enables mechanical ventilation. However, a distally placed endotracheal tube may obscure proximal tracheal lesions or stent malacic tracheal segments.

FURTHER READING

Ayari-Khalfallah S, Froehlich S. Airway endoscopy and assessment in children. In: Graham JM, Scadding GK, Bull PD (eds). *Pediatric ENT*. Berlin: Springer; 2007: pp. 177–82.

Casselbrant ML, Alper CM. Methods of examination. In: Bluestone CD, Stool SE, Alper CM, et al. (eds). *Pediatric Otolaryngology*. Philadelphia, PA: Saunders; 2003: pp. 1379–94.

Green CG, Holinger LD, Gartlan MG. Technique. In: Holinger LD, Lusk RP, Green CG (eds). *Pediatric Laryngology and Bronchoesophagology*. Philadelphia, PA: Lippincott-Raven; 1997: pp. 97–116.

Kuo M, Rothera M. Emergency management of the pediatric airway. In: Graham JM, Scadding GK, Bull PD (eds). *Pediatric ENT*. Berlin: Springer; 2007: pp. 183–8.

Mani N, Soam M, Massey S, et al. Removal of inhaled foreign bodies – middle of the night or the next morning? *Int J Pediatr Otorhinolaryngol* 2009; 73: 1085–9.

Oncel M, Sunam GS, Ceran S. Tracheobronchial aspiration of foreign bodies and rigid bronchoscopy in children. *Pediatr Int* 2012; 54(4): 532–5.

Soyer T. The role bronchoscopy in the diagnosis of airway disease in children. *J Thorac Dis* 2016; 8(11): 3420–6.

5

Esophageal dilatation

JENNIFER BILLINGTON AND NIYI ADE-AJAYI

HISTORY

Interventions for children with esophageal strictures have been performed since the Middle Ages. Fabricius ab Aquapendente first described instrumentation of the esophagus with tapered lengths of wax in the 16th century. The term "bougienage" derives from the French word for a wax candle, a *bougie*. Other pioneers such as Ambrose Pare (1510–1590), Wilhem Fabry (1560–1634) and Thomas Willis (1621–1675) carried out disimpaction and dilatation of the esophagus with readily available natural materials including leather tubes, osiers made from willow, whale bone, and swan quills. A number of descriptive phrases were used early on including esophageal "sounds," a term still used in modern medicine. This was thought to originate from the noise of esophageal dilators hitting impacted, metal foreign bodies. These original dilators were the precursors of a range of manufactured and more effective ones such as the gum elastic bougies developed by the American laryngologist Chevalier Jackson (1865–1958).

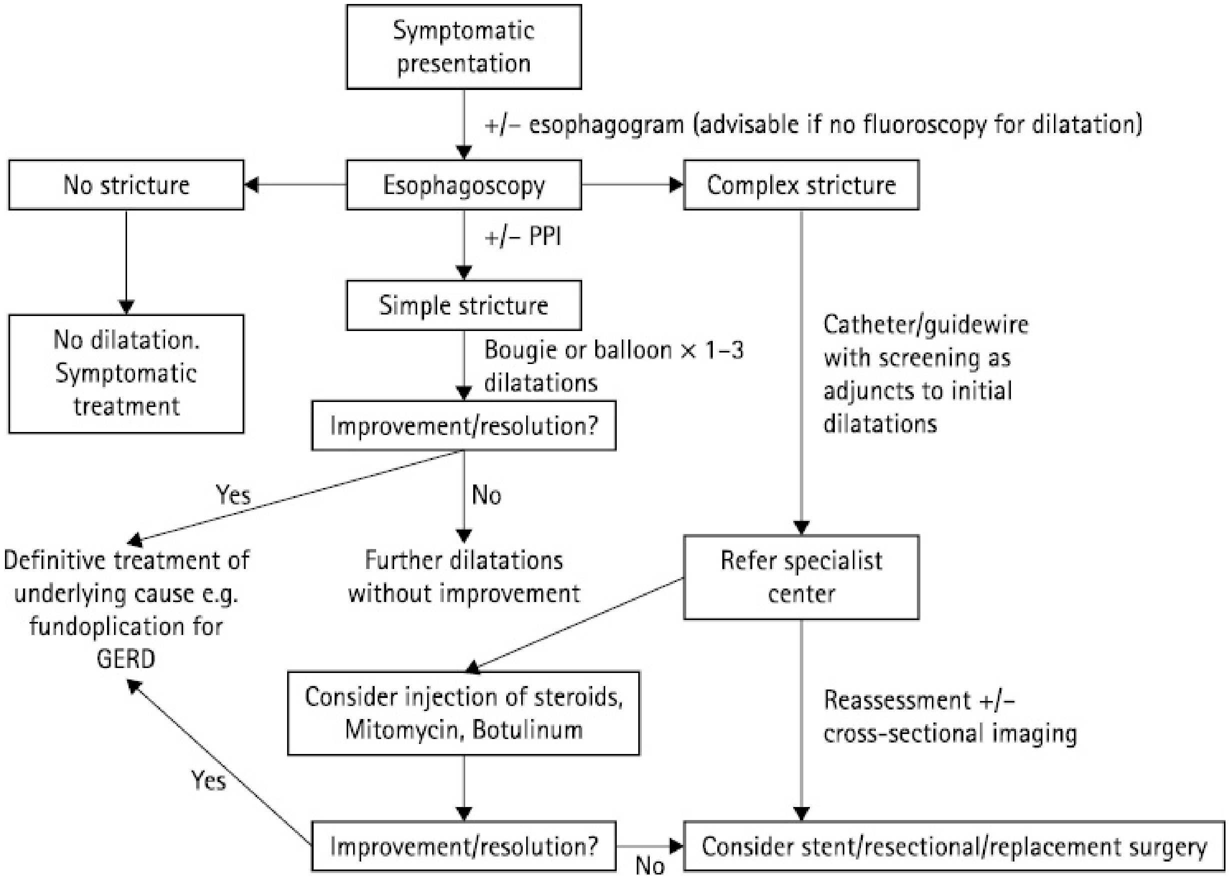
Techniques have evolved over time with rigid (1850s), semi-rigid (1970s), and more recently flexible esophagoscopy. Although dilatation has been proven an effective technique by direct, largely blind, instrumentation, in due course with the advent of endoscopy and now radiological imaging, other techniques have largely superseded it.

PRINCIPLES AND JUSTIFICATION

The vast majority of affected children are affected by benign disease. Anastomotic stricture following esophageal atresia repair is one of the most common causes followed by gastroesophageal reflux disease (GERD) and reflux-associated stricture. Less commonly, causes include caustic ingestion, achalasia, cartilaginous rings, and congenital esophageal stenosis may be seen (Table 5.1 and Figure 5.1).

Table 5.1 **The aetiology of esophageal obstruction**

Category	Condition	Types/characteristics
Congenital	Muscular hypertrophy Esophageal web Cartilaginous ring	Obstruction is typically in distal esophagus
Acquired	GERD	Peptic, short, few dilatations if early; recalcitrant if long-standing and fixed
	Corrosive ingestion	Caustic, irregular, often long, tend to be recalcitrant
	Postoperative	Anastomotic stricture postesophageal atresia repair, tight fundoplication, sclerotherapy, bands
	Achalasia	Needs large balloon to overdilate
	Schatzki's ring	Complete mucosal ring usually above the gastroesophageal junction occurring predominantly in adolescents Aim to disrupt ring at dilatation and treat with PPIs
	Epidermolysis bullosa	Shearing forces should be avoided during dilatation Balloon superior to bougie
	Foreign body	Fixed if long-standing Beware tracheoesophageal fistula


Symptomatic presentation
+/– esophagogram (advisable if no fluoroscopy for dilatation)
No stricture
Esophagoscopy
Complex stricture
+/– PPI
Simple stricture
No dilatation. Symptomatic treatment
Bougie or balloon × 1–3 dilatations
Catheter/guidewire with screening as adjuncts to initial dilatations
Improvement/resolution?
Yes
No
Definitive treatment of underlying cause e.g. fundoplication for GERD
Further dilatations without improvement
Refer specialist center
Consider injection of steroids, Mitomycin, Botulinum
Reassessment +/– cross-sectional imaging
Yes
Improvement/resolution?
No
Consider stent/resectional/replacement surgery

5.1 Algorithm for esophageal dilatation. PPI, proton pump inhibitor; GERD, gastroesophageal reflux disease.

Children at risk of developing strictures should be closely monitored clinically and with the judicious use of radiology and endoscopy so that, when indicated, intervention is timely. Some children put forward for dilatation may be more appropriately managed conservatively with, for example, proton pump inhibitors (PPIs). Others may be more suited to bypass by way of gastrostomy while others may require stricture resection or even esophageal replacement surgery.

It is useful to consider strictures in terms of complexity. Simple strictures are short and straight while complex ones are tight, long, and irregular. The majority of strictures are amenable to dilatation using one or more of the methods described below. However, recent perforation is a contraindication. The key goals of intervention are the relief of dysphagia, the achievement of effective oral nutrition, and airway protection. Oral aversion is a notable difficulty for those with recalcitrant structures, thus early and regular intervention is essential in order to maximize the potential for feeding. At the commencement of intervention, an estimate of the number of dilatations likely to result in cure is helpful. This helps to avoid children having large numbers of dilatations without commensurate clinical benefit. Those with complex esophageal strictures who have undergone numerous dilatations with little improvement in dysphagia and limited oral nutritional intake are best managed in the context of a multidisciplinary team, including surgeon with esophageal replacement expertise, gastroenterologist, interventional radiologist, and dietician. When the multidisciplinary expertise is not locally available, referral to an appropriate center is advisable.

OVERVIEW OF DEVICES AND TECHNIQUES

The range of available procedures has increased over the past few decades. The choice of device and specific technique is dependent on the condition being treated and characteristics of the stricture. While a specific plan for dilating should be made preoperatively, a wide range of tools should be available. An esophageal contrast study may be helpful at the outset, particularly if fluoroscopy is not available for dilatation. Flexible endoscopic assessment is recommended, although rigid endoscopy also allows for direct visualization and remains a suitable alternative. At the outset, detailed information regarding the stricture should be documented and include site, degree of tightness, length, and mucosal inflammation.

Devices for dilatation may be put into three broad categories: Bougies, balloon dilators, and stents. Bougies can be subdivided into those that are passed over a guidewire and those that are not. They come with a range of different tips: Blunt, conical, olive-tipped, and tapered. Stents are divided depending on material (metal or plastic) and whether or not they are covered (**Tables 5.2 and 5.3**).

Table 5.2 Conversion chart

Conversion chart (diameter = Fr/3)	
French (Fr or Fg) (Charrière or Ch)	**Millimeters (outer diameter)**
2	0.7
6	2.0
10	3.3
15	5.0
20	6.7
25	8.8
30	10.0
35	11.7
40	13.3
50	16.7
55	18.3
60	20.0

Table 5.3 Dilators

Subtype of dilator	Material	Characteristics	Size/increments	Advantages	Disadvantages
Bougies					
Savary–Gilliard	Polyvinyl chloride (PVC)	Long and tapered	3 Fr increments (max. 60 Fr)	Reusable Widely available Radiological marker	Instrument maintenance, especially microbiological
Eder–Puestow	Metal	Central hollow for guidewire Graduated olive shape	2 Fr increments (max. 51 Fr)	Flexible Reusable	Oral trauma
Chevalier–Jackson	Polythene, gum elastic	Passed via rigid esophagoscope	Max. 60 Fr	Have memory when curved into shape	Perform best when warmed
Filiform	Polythene, gum elastic	Fine filiform is inserted via stricture then a “follower” is attached and used to achieve dilatation	Filiform 2–6 Fr Follower 10–24 Fr	Fine tip allows navigation through narrow strictures	Size range
Tuckers	Silicone	Loop each end to tie to string	2 Fr increments (max. 60 Fr)	Suitable for dilatation of long-standing strictures	
Maloney	Rubber–mercury- or tungsten-filled	Tapered tip May be used blindly, but fluoroscopic control recommended	2 Fr increments (max. 60 Fr)	Gentle taper Can be used with/out fluoroscopy	Mercury dilators being replaced over concerns about mercury poisoning
Balloons					
Balloon	Thermoplastic polymers	Non-latex balloons passed deflated down the side port of endoscope under vision + filled with radiopaque solution	Single or multiple diameter balloons (max. 60 Fr)	Accurate fluoroscopic positioning across narrowest point Direct visualization	Cost Disposable

				Expanded with/out pressure monitor
Special achalasia balloons	Polythene	Wire-guided, large diameter	90–135 Fr	Large enough to achieve disruption of lower esophageal muscle fibers

The rigid endoscope allows direct assessment of the stricture and facilitates the passage of smaller bougies. Flexible endoscopy enables direct visualization and in addition allows the targeted introduction of a guidewire or balloon. The endoscope can then be withdrawn and be replaced with an appropriate bougie or balloon dilator. If a guidewire is used, insertion can be achieved fluoroscopically using a catheter. A number of catheter types and guidewires may be suitable, but typically for a tight stricture, a floppy-tipped, hydrophilic guidewire (e.g. 0.035 inch) is passed via a 5 Fr catheter with curved tip. Screening contrast after dilatation need not be routine, particularly for simple strictures. However, the threshold for carrying this out immediately post-dilatation or subsequently should be low for early diagnosis of a perforation and reduced morbidity. If early repeat dilatation of a tight stricture is planned, a nasogastric tube with the tip cut off may be left in place to facilitate the next procedure.

While there are many advantages to reusable dilators, stringent antimicrobial policies in many hospitals have led to a decline in the use of these devices and a greater reliance on disposable dilators, particularly balloons.

PREOPERATIVE

Assessment and planning

Early classification of whether a child has an acquired or congenital structure often prompts an appropriate treatment pathway.

Delineation of the esophageal anatomy is a fundamental step when deciding an appropriate treatment pathway. An upper gastrointestinal (GI) contrast study may yield information about the stricture position, regularity, and length. Establishing whether there is evidence of external compression along with associated conditions such as reflux and hiatus hernia may be valuable.

For many patients, planning both the number and frequency of dilatations is on a case-by-case basis. For example, patients with a long-standing post-anastomotic stricture, small incremental dilatations may be ideal, whereas for achalasia a more forceful dilatation with the aim of disrupting the fibers of the distal esophageal sphincter is appropriate.

Similarly, for those who require multiple dilatations, frequency between dilatations should be carefully considered. Early on in the series of interventions, the intervals may necessarily be of short duration. The intervals can then be lengthened as a pattern is established. Dilatation of simple strictures or those that have previously been dilated without difficulty can be planned on a day-case basis.

Esophageal dilatations may be carried out in an operating theatre, in a radiology department, or in specially designed suites with interventional radiology guidance. Routine antibiotics are recommended for infants and small children and should be used selectively for those with long-established strictures or other indications such as cardiac anomalies.

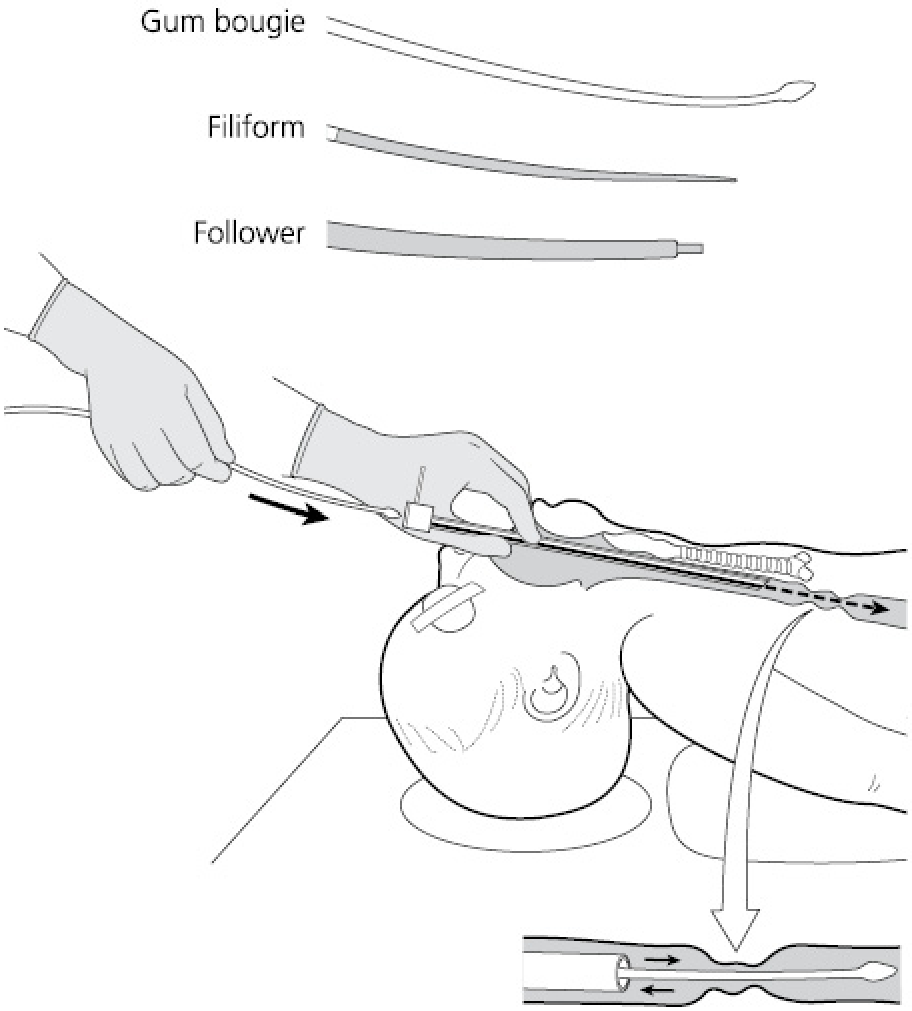
Anesthesia

General anesthesia with endotracheal tube intubation is our preferred method for children with an esophageal stricture. In selected cases, such as those with short strictures requiring frequent dilatations, sedation may be considered. However, sedation is poorly tolerated by many children and may compromise the effectiveness of the intervention.

OPERATIONS

Bouginage of esophageal stricture without guidewire

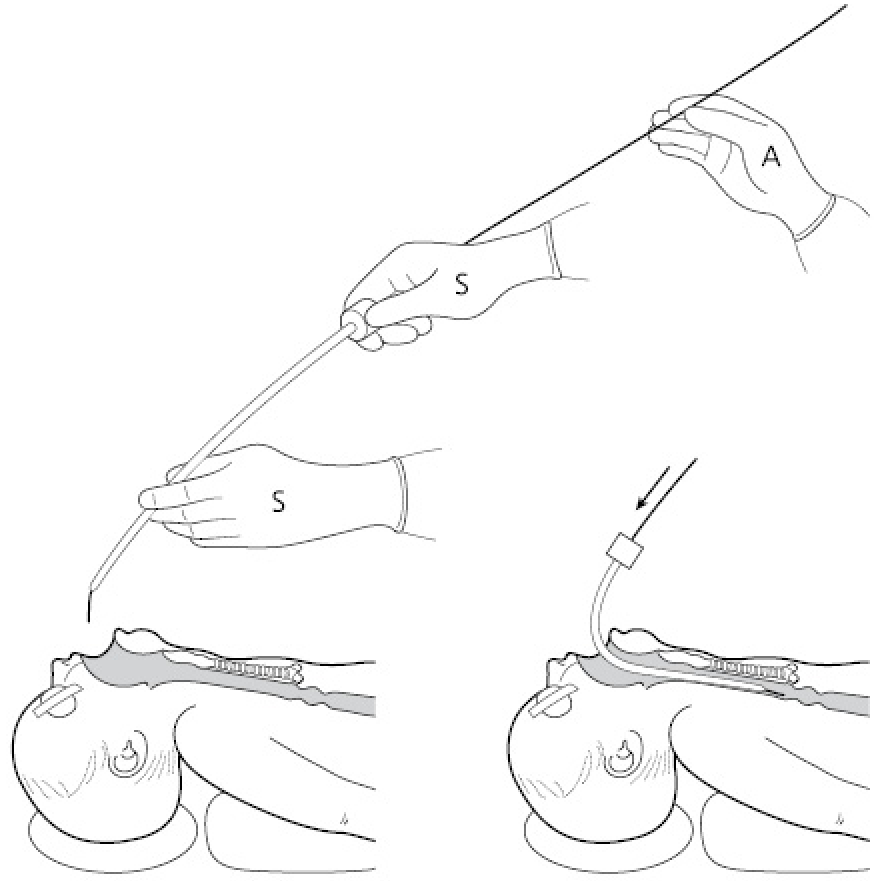
This procedure is preceded by rigid esophagoscopy with the child in the supine position and the neck extended on a head ring (**Figure 5.2**). The index finger of the operator's non-dominant hand is used to pull back the upper teeth and the thumb used to protect the teeth from damage by the esophagoscope. With the esophagoscope advanced to immediately proximal to the stricture, the selected bougie is prepared with hydrophilic gel and gently passed. The ideal grip involves thumb and index finger, supported behind by the middle finger; a gentle rather than a "fist" grip. This is preferred in order to reduce the amount of force applied and thus lessen the risk of perforation. The choice of initial dilator size is based on radiological/endoscopic assessment. Blind bouginage, particularly for first time dilatation, is not recommended.



5.2

Bouginage of esophageal stricture with guidewire

A suitably shaped catheter is inserted into the upper esophagus with fluoroscopic guidance and a contrast study is performed. A stricture is demonstrated and is then crossed with a guidewire, which is then advanced to the stomach. The combination of a curved (steerable) tip of the catheter and fine floppy guidewire ensures that the majority of even very tight strictures can be traversed. The guidewire channel of a suitably sized and shaped bougie (e.g. Savary–Gilliard®) is irrigated and advanced over the guidewire. Dilatation is carried out progressively as appropriate (**Figure 5.3**).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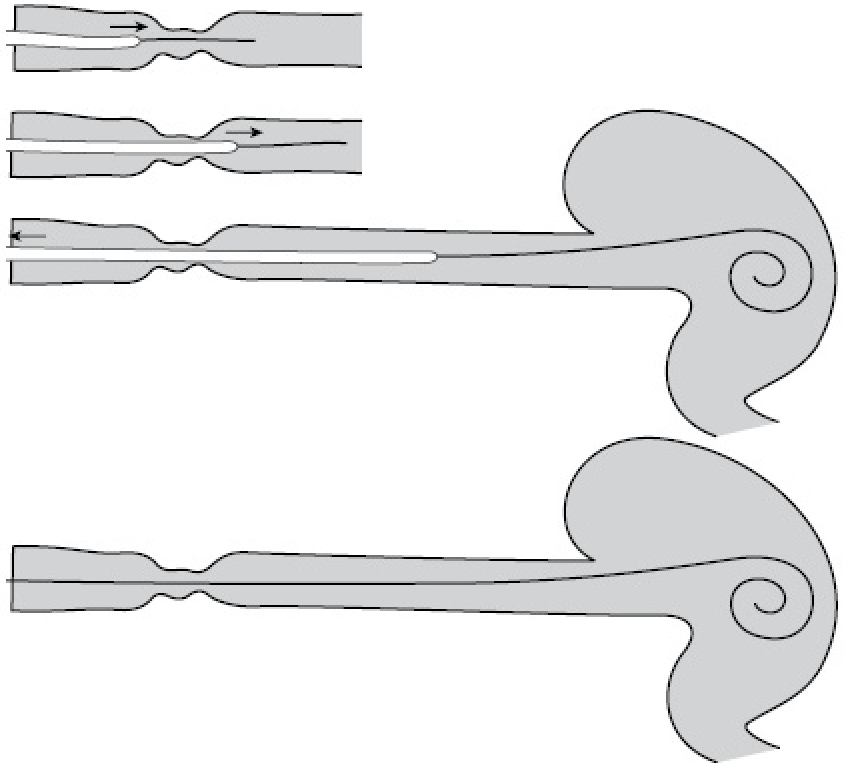
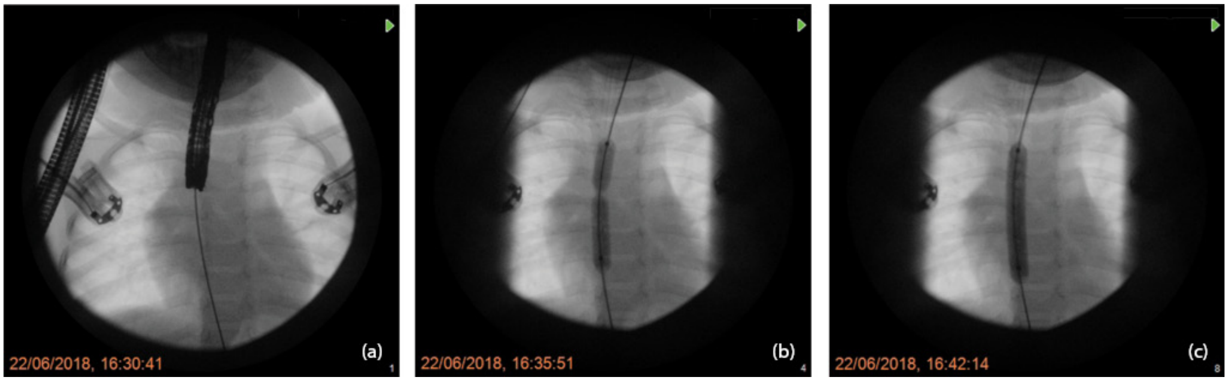


5.3

Balloon dilatation under fluoroscopic control

For practical purposes, fluoroscopic and endoscopic balloon techniques are often interchanged to achieve optimum outcomes. Balloon size is determined by the estimated size of the normal esophagus of the child, tightness and duration of stricture, and the underlying condition. An early peptic stricture, even when tight, should dilate up readily, and balloon size could be taken from the size of the child's thumb. On the other hand, a caustic stricture should be dilated in small increments. If the underlying condition is achalasia, the aim is to dilate by rupturing and a large balloon size should be chosen.

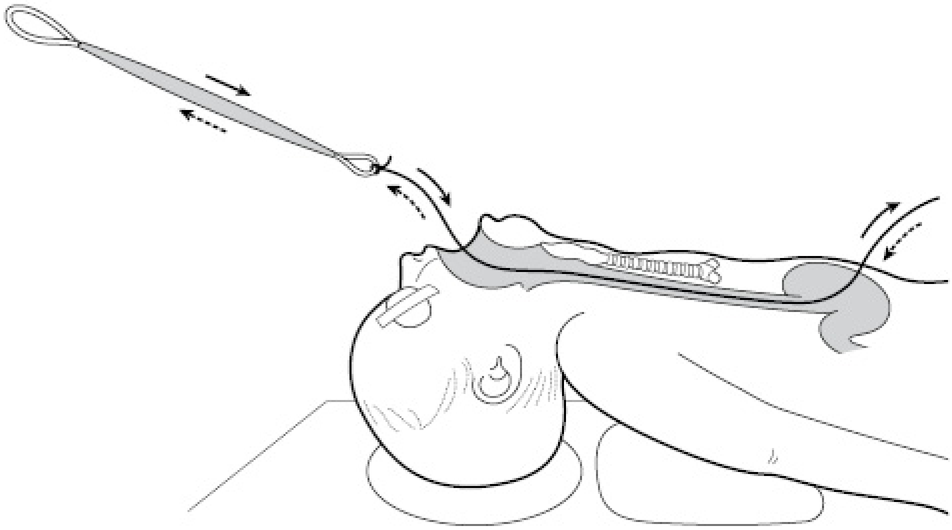
The flexible endoscope is passed as outlined elsewhere in this text until it is just proximal to the stricture (**Figure 5.4a**). The instrument channel should be irrigated with saline. A suitable wet guidewire with a floppy tip (typically 0.035 inch and 260 cm) is advanced via the instrument channel under direct vision. Redundant guidewire in the stomach should be confirmed fluoroscopically. The endoscope is then slowly withdrawn, ensuring that, as this takes place, commensurate insertion of the guidewire occurs to maintain the original position in the stomach. As the endoscope is removed, the guidewire is controlled at the level of the lips to allow complete disengagement of the endoscope from the guidewire without dislodging it. The angioplasty catheter is advanced over the guidewire until the radio-opaque markers indicating the position of the balloon straddle the stricture (**Figure 5.4b**). The balloon is then inflated with dilute radiographic contrast (**Figure 5.4c**) with the aid of a pressure monitor. Fluoroscopy is essential at this stage to confirm that the stricture has been successfully dilated, as shown by abolition of the waist on the balloon. There is no general agreement on the optimum duration of dilatation, but prolonged maintenance of pressure does not appear to improve the results. Our practice is to maintain pressure for 1–3 minutes once the desired diameter has been reached. The balloon is then deflated and the catheter and guidewire are removed. There is often a small amount of blood on the dilator. In the main, this does not pose a clinical problem. Stricture position and the starting and completion diameter should all be carefully recorded, including relationship with bony and soft tissue landmarks.

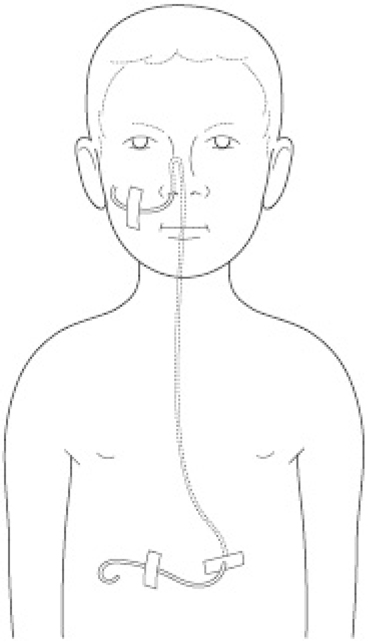


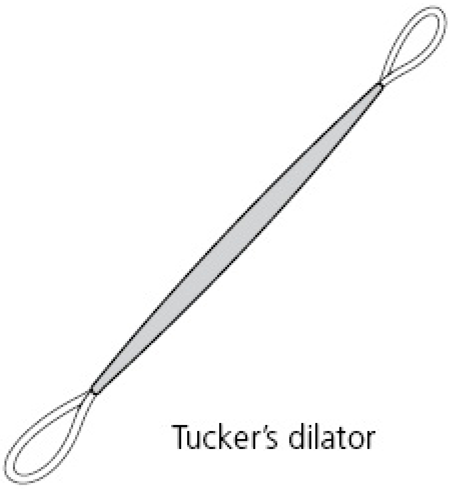
5.4a–c Chest radiographs showing balloon dilatation: (a) Before, (b) during, and (c) after.

Retrograde dilatation

If antegrade cannulation of a tight stricture proves impossible, it can be approached from below. This is usually carried out via an established gastrostomy and can be facilitated by flexible endoscopy, or fluoroscopy and guidewire (**Figure 5.5**). Once the stricture has been traversed, every effort should be made to keep the lumen open and a channel for future rapid dilatation. A nasogastric tube between dilatations usually achieves this. The tip should be cut off in order to be able to pass the guidewire and retrieve the nasogastric tube during the next dilatation.







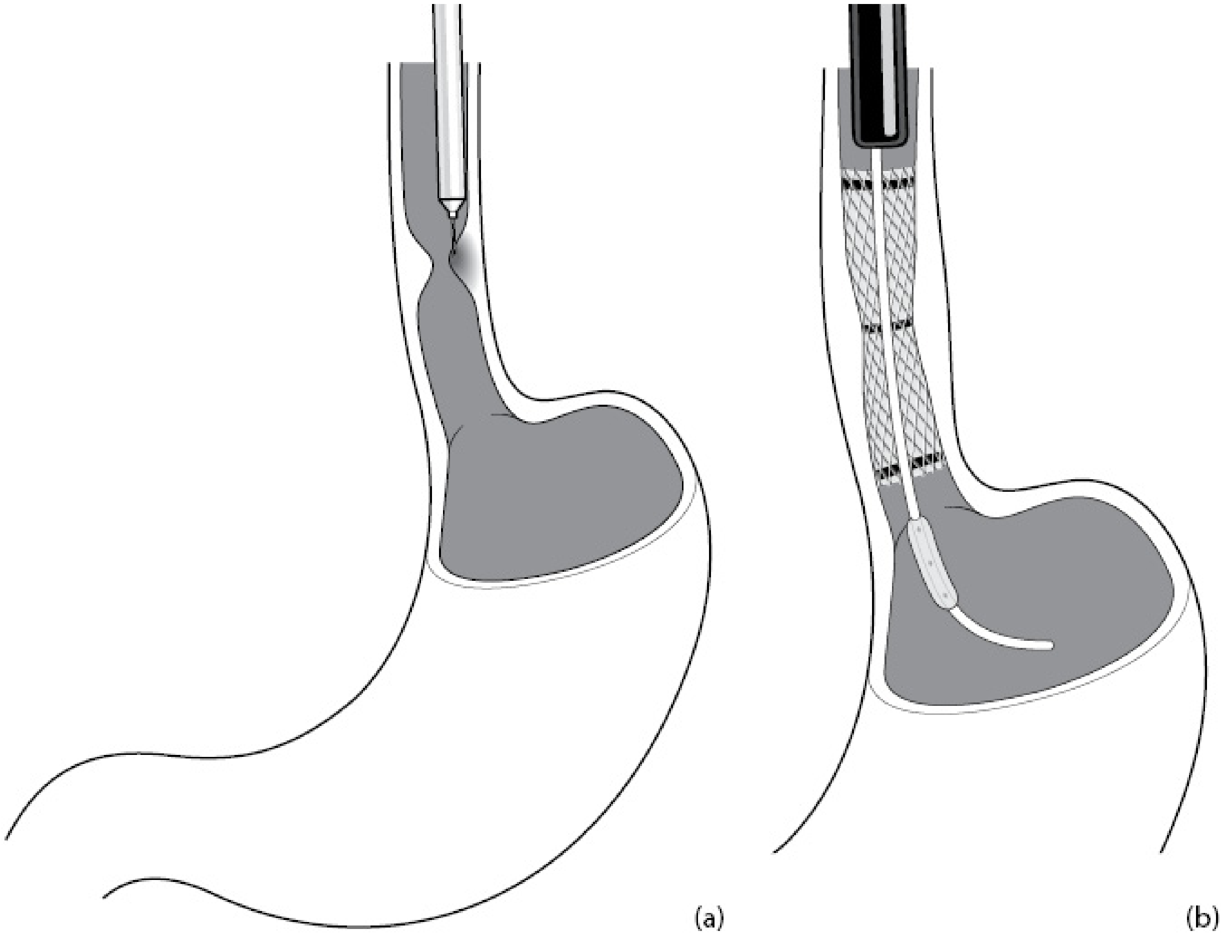
5.5

Alternative strategies for recalcitrant strictures

When multiple conventional dilatations have failed to achieve sustained luminal opening and relief of symptoms, alternative methods, usually in the context of a specialist center are appropriate. These include direct endoscopic injection into the stricture. Agents that can be injected include steroids, mitomycin, and botulinum toxin. A four-quadrant technique is recommended, with the overall dose divided into four and administered into each quadrant.

When maximal efforts to preserve the esophageal lumen fail to resolve symptoms, stenting may be an option. There are currently a broad range of stents (e.g. metal, plastic, polymerized, and biodegradable materials) that may represent a medium-term solution for persistent dysphagia. These may be uncovered, fully covered, and partially covered. The recent introduction of biodegradable stents, which promote tissue ingrowth and do not need to be retrieved, may provide an attractive treatment option, particularly in caustic stricture patients.

Stents come in a variety of lengths and diameters in order to best suit the anatomy of the individual child. Some cases require "made-to-measure" stents. Regardless of stent, the principles remain the same; the stent is deployed over a wire with direct endoscopic and fluoroscopic guidance with the aim of traversing the stricture and ensuring that the mid portion of the stent sits at the narrowest point (**Figure 5.6a and b**). Conventional dilatation of the stricture and passage of a trans-stricture guidewire under general anesthesia should precede stent insertion. When the correct position has been confirmed fluoroscopically, the stent is deployed.



5.6a,b

Esophageal stent insertion has a higher complication rate compared with dilatation alone. Stents are therefore reserved for children for whom more traditional methods have failed. Postoperative complications can be classified as early (≤4 weeks) and delayed (>4 weeks). Early complications include bleeding, gastroesophageal reflux, stent intolerance, perforation and migration. Delayed complications include all the early ones as well as stent occlusion and formation of fistulas into nearby structures.

POSTOPERATIVE CARE

If a nasogastric tube has been left in place, this should be secured with occlusive, transparent dressing. Monitoring following dilatation should be carried out in a designated recovery area immediately following dilatation and should include regular measurements of pulse, heart rate, temperature, and blood pressure. If an adequate luminal diameter has been achieved, clear fluids may be permitted by mouth when the child is fully awake. If these are tolerated, then progression to milk is allowed followed by a soft diet if appropriate.

TREATMENT OF STRICTURES IN LOW- AND MIDDLE-INCOME COUNTRIES

In low- and middle-income countries (LMICs), the disease burden and etiologies of esophageal strictures are somewhat different. While the incidence of esophageal atresia (EA) and tracheoesophageal fistula (TEF) is broadly comparable with developed countries, the incidence of esophageal injuries secondary to caustic substance ingestion is considerably higher. Many children have access to caustic substances at home and in workplaces, and a general lack of appropriate packaging and healthcare labels results in frequent cases of corrosive esophageal damage.

Difficulty in accessing appropriate healthcare resources, widespread poverty, and lack of funding all continue to affect the delivery of care to patients with esophageal strictures. Children will present with common symptoms of esophageal obstruction including weight loss, malnutrition, food bolus obstruction, and aspiration events. For those affected by caustic strictures, attempts to dilate are encouraged, with most centers reporting success rates in excess of 60%. Along with an increased morbidity, there is additionally an increased likelihood of requiring repeated dilatations when compared with other stricture etiologies in children.

Availability of resources depends to a considerable extent on location, with some centers offering balloon dilatation while others struggle to provide the most basic of bougies. Equipment availability, cleaning, and upkeep costs are a rate-limiting step for resource-depleted regions. In some centers, prograde dilatation is described with or without a guidewire trans-stricture string. The placement of a nasogastric tube following dilatation with the tip removed not only permits passage of a wire for future dilatation options but also can allow for enteral feeding. This may be useful in identifying the esophageal lumen in severe strictures, particularly those affecting the pharynx. However, as the scope is passed orally, identification of the tube trajectory from the nose downwards may be challenging.

In order to promote normal mastication and oropharyngeal functioning, surgical replacement of the native eosphagus should be considered after multiple failed dilatations. Options include interposition with stomach, jejunum, or colon.

OUTCOME

The majority of children with an esophageal stricture have a satisfactory and sustained response to a few dilatations. In patients with EA/TEF strictures, balloon dilatation is successful in 90% of patients, with half requiring two or more dilatations to achieve complete resolution of symptoms. Complication rates are low in experienced hands. They occur most often during dilatation of corrosive strictures followed by those associated with GERD. Other at-risk patients include those with dystrophic epidermolysis bullosa. Careful postoperative monitoring and a low threshold for chest X-ray and upper GI contrast study facilitate early diagnosis. Three types of esophageal injury occur. Guidewire perforation of the esophagus can be recognized at fluoroscopy and treated conservatively. Submucosal tears of the esophagus are seen as contained extravasation of contrast. These also seem to do well with conservative treatment. Full-thickness perforation is uncommon. Small perforations require nasogastric or parenteral feeding and broad-spectrum antibiotic therapy. Large perforations, if recognized early, may be managed best by surgical repair.

CONCLUSION

While the etiology of esophageal strictures is different in children from that in adult patients, the goal is the same—to establish a normal esophageal lumen which alleviates symptoms and promotes feeding. Knowledge of the anatomy of each individual case enables the endoscopist to make the best choice of treatment plan and interval planning. Although no clear difference has been established between the effectiveness of bougie and balloon dilatators, microbiological considerations and the appeal of circumferential dilatation make balloons increasingly attractive in the high-income setting.

FURTHER READING

Arnold M, Numanoglu A. Caustic ingestion in children – a review. *Semin Pediatr Surg* 2017; 26: 95–104.

Berger M, Ure B, Lacher M. Mitomycin C in the therapy of recurrent esophageal strictures: Hype or hope? *Eur J Pediatr Surg* 2012; 22: 109–16.

Earlam R, Cunha-Melo JR. Benign oesophageal strictures: Historical and technical aspects of dilatation. *Br J Surg* 1981; 68: 829–36.

Fallon BP, Overman RE, Geiger JD, et al. Efficacy and risk profile of self-expandable stents in the management of pediatric esophageal pathology. *J Pediatr Surg* 2019; 54: 1233–8.

Lan LCL, Wong KKY, Lin SCL, et al. Endoscopic balloon dilatation of esophageal strictures in infants and children: 17 years' experience and a literature review. *J Pediatr Surg* 2003; 38: 1712–15.

Lew RJ, Kochman ML. A review of endoscopic methods of esophageal dilation. *J Clin Gastroenterol* 2002; 35: 117–26.

Pieczarkowski S, Woynarowski M, Piotr L., et al. Endoscopic therapy of oesophageal strictures in children – a multicentre study. *Prz Gastroenterol* 2006; 11: 194–9.

6

Esophagogastroduodenoscopy and endoscopic retrograde cholangiopancreatography

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HISTORY

Endoscopists in the 19th century used open, rigid tubes to visualize the upper gastrointestinal (GI) tract. The quality of rigid endoscopes was improved by the addition of conventional lenses that provided magnification and increased viewing angles. Flexible fiberoptic endoscopes were first developed in 1958. Flexible endoscopes for upper GI endoscopy are now available in small sizes ideally suited to pediatric applications.

Technical improvements and the development of specialized equipment have expanded the diagnostic and therapeutic potential of endoscopy. Introduced in adults in 1968 and children in 1976, endoscopic retrograde cholangiopancreatography (ERCP) is an integral tool in the evaluation and management of many biliary and pancreatic diseases. New technology and techniques including endoscopic ultrasound and double balloon enteroscopy have allowed for evaluation of pathologies previously thought unreachable by endoscopy. Endoscopy has also played an increasing role in minimally invasive surgery ("endosurgery" or "endolumenal surgery"), assisting – and in some cases replacing – thoracoscopic and laparoscopic approaches.

ESOPHAGOGASTRODUODENOSCOPY

PRINCIPLES AND JUSTIFICATION

Indications

Endoscopy of the upper GI tract is performed for both diagnostic and therapeutic reasons:

- Upper GI tract bleeding
- Ingested foreign bodies
- Caustic ingestion
- Esophageal injury
- Esophageal stricture and achalasia
- Reflux disease
- Tracheoesophageal fistula evaluation
- Evaluation of dysphagia, pain, and bleeding

Owing to increased awareness of diseases including Crohn's disease, eosinophilic esophagitis, and celiac disease, the incidence of first-time esophagogastroduodenoscopy (EGD) use in the pediatric population has grown greater than 12-fold in the last 20 years. Specific indications for additional therapeutic modalities of endoscopy are discussed below.

UPPER GI TRACT BLEEDING

Flexible EGD is used in the evaluation and management of upper GI tract bleeding (proximal to the ligament of Treitz). Definitive or palliative treatment may be provided for specific lesions, and localization of the bleeding source is critical in the event of refractory bleeding warranting surgical intervention. In severe cases, proper resuscitation to achieve hemodynamic stability prior to endoscopy is essential. Esophageal varices, caused by portal hypertension, may be treated by endoscopic sclerotherapy or variceal banding. Most upper GI bleeding in children is non-variceal, caused by peptic ulcers, erosive esophagitis, Mallory–Weiss tears, and vascular malformations. Ulcer bleeding may be controlled by injection of epinephrine, mechanical therapy with clips, and coagulation using bipolar or monopolar electric current or heat.

INGESTED FOREIGN BODIES

Either flexible or rigid esophagoscopy is mandatory in the evaluation and treatment of suspected esophageal foreign bodies. When encountered, foreign bodies may be removed or advanced into the stomach. Most gastric foreign bodies will pass spontaneously. Those that fail to pass after a period of observation require extraction with a flexible scope. Accidental ingestion of magnets or button batteries is increasing in incidence, has grave implications, and is discussed in detail below.

CAUSTIC INGESTION/ESOPHAGEAL INJURY

Esophagogastroscopy is useful in the evaluation of infants and children following suspected ingestion of caustic substances as clinical manifestations (including symptom severity and the presence of oral lesion) poorly correlate with esophageal damage. The degree of esophageal injury, although difficult to assess, remains the best predictor of morbidity and mortality. The likelihood of subsequent stricture formation is also difficult to determine from the appearance of the mucosa. Follow-up evaluations can assess mucosal healing and early stricture formation. As most children have mild injury after caustic ingestion, the role of mandatory esophagoscopy in all caustic injury patients is debated.

ESOPHAGEAL STRICTURE AND ACHALASIA

Endoscopy is beneficial in the diagnosis and treatment of strictures and achalasia. Strictures encountered may result from:

- Peptic disease or reflux
- Congenital cartilaginous bands
- Eosinophilic esophagitis
- Postoperative stricture of the esophagus

Esophageal dilation is performed using prograde sequential dilators, with or without guidewires, and balloon dilators. Large-caliber balloon dilation is used to treat achalasia. Fluoroscopy to confirm proper guidewire position and to monitor the passage of each dilator is a useful adjunct for more complex strictures; however, it may not reduce the risk of perforation. Traditionally, mechanical (bougie) dilators of progressively larger size are passed to the point of maximal dilation. Clear end-points for dilation are difficult to determine (e.g. blood on dilator, subjective increase in difficulty). Balloon dilators apply force to the stricture in a radial fashion and may be safer than mechanical dilators that have both radial and longitudinal force vectors. Esophageal injury and perforation can occur with any technique. Strictures refractory to dilation may require additional endoscopic management including intralesional steroid or mitomycin C injection, stent placement, and endoscopic incisional therapy.

ADJUNCTS TO FEEDING

Endoscopic techniques may be used to place gastrostomy tubes (percutaneous endoscopic gastrostomy) and transpyloric feeding tubes (see Chapter 34).

CHOICE OF ENDOSCOPE

The choice of rigid or flexible endoscopes depends on the procedure planned and the experience and training of the surgeon. The safety and ease of flexible endoscopy make it the procedure of choice for most pediatric upper GI endoscopic procedures. The ability to insufflate air and thus distend the esophagus and allow complete visualization ahead of the advancing esophagoscope is the principal advantage of the fiberoptic endoscope. Endoscopists should be familiar with both techniques.

Rigid esophagoscopes

The Storz–Hopkins rod-lens telescope is utilized almost exclusively in rigid esophagoscopy. Unlike a conventional telescope that uses small lenses with long intervening air spaces, this system uses long glass rods with their ends shaped in the form of a lens and small intervening air space. The lens system allows transmission of a brilliant magnified image through a small-diameter tube. A wide variety of endoscopic instruments is available to perform such tasks as foreign body retrieval and sclerotherapy. The rigid scope can also be used to “sheath” foreign bodies, allowing for safer extraction of sharp objects. General anesthesia with endotracheal intubation is required.

Flexible endoscopes

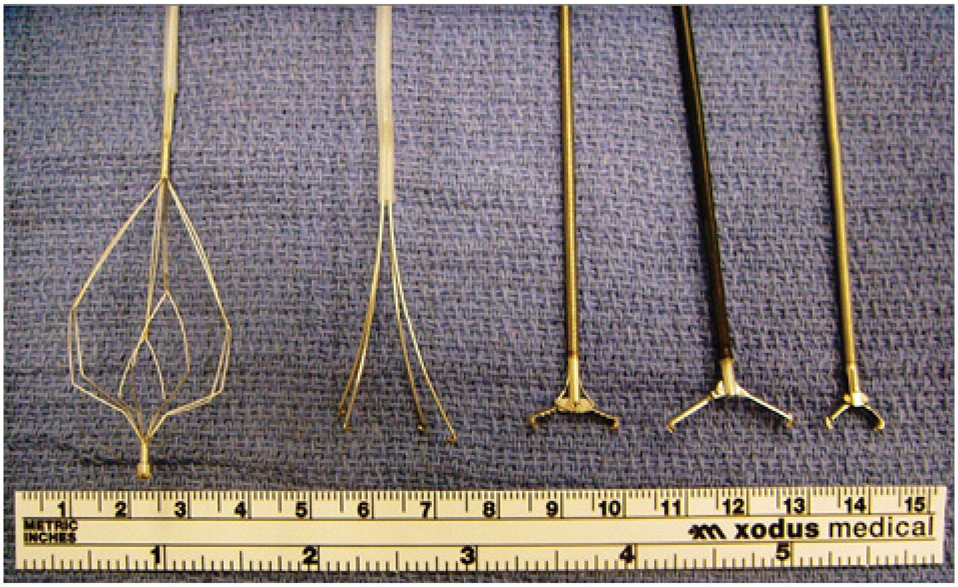
The surgeon must be aware of the length, diameter, working channel size, and types of accessories needed (as well as their compatibility with the selected endoscope) prior to beginning any flexible endoscopic intervention (**Table 6.1**).

Table 6.1 **Flexible endoscope options**

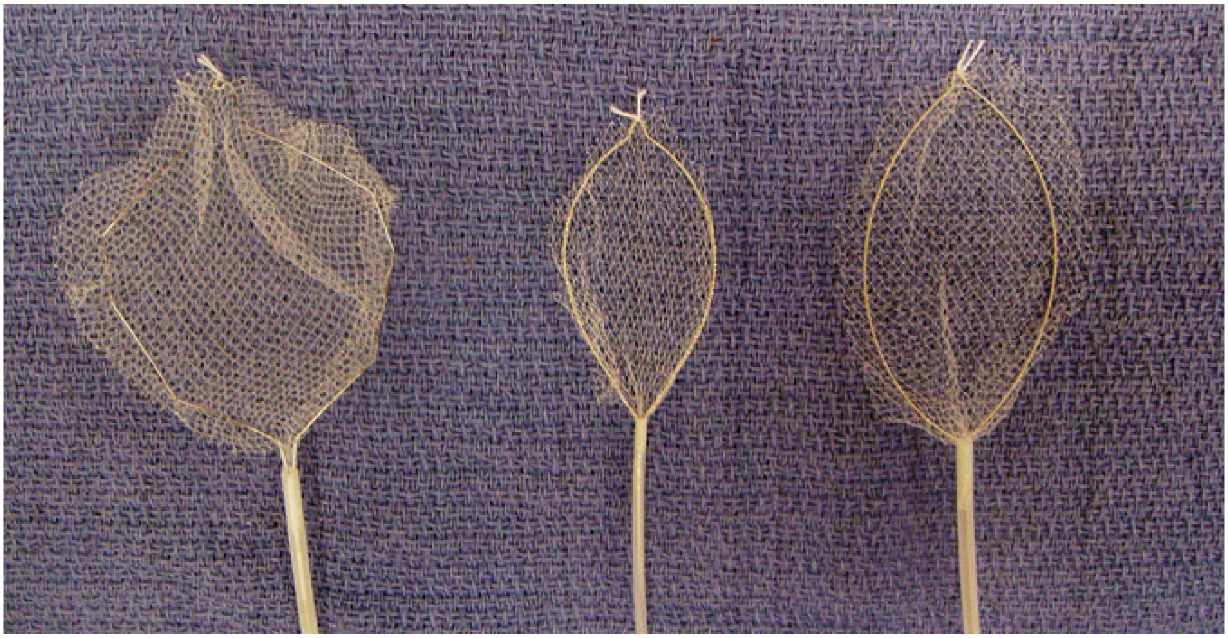
Diameter (mm)	**Working channel diameter (mm)**	**Advantages**	**Disadvantages**
4.9–5.4	2.0–2.2	Can be used for smaller children or patients with high-grade strictures	Requires dedicated small-caliber endoscope accessories
8.0–11.4	2.8–3.7	“Standard” adult-sized endoscope: • Permits intragastric retroflexion for fundic visualization or transduodenal passage • Vast majority of endoscopic accessories fit through channel	Only one accessory channel Too large for some children
12.5	2.8 and 3.7 (two channels)	Two accessory channels can aid in specific therapeutic maneuvers and permit specific devices	Outer diameter limits usefulness in younger children

Endoscopic accessories

The therapeutic potential for endoscopy is broadened by continual technological advances in endoscopic accessories. A wide array of foreign body graspers and snares are available for foreign body removal (**Figure 6.1**). Retrieval nets and baskets in various sizes and shapes can be chosen based upon the size and location of the foreign body (**Figure 6.2**). Nets and baskets are particularly valuable for removing round foreign bodies that can be difficult to hold with graspers, such as marbles and ball bearings. Sharp objects (e.g. glass, bones, razor blades) can be safely removed using a wide variety of foreign body hoods and overtubes. Softer objects such as meat impactions may be more easily removed with suction catheters.



6.1 Examples of foreign body graspers.



6.2 Examples of foreign body retrieval devices.

Biopsy techniques

Small cup biopsy forceps that pass easily through the instrument channel of the flexible endoscope or alongside the telescope within the rigid endoscope allow precisely directed biopsies to be performed. Additional biopsy techniques include both suction and needle biopsy.

PREOPERATIVE

GI endoscopy should be performed either in the operating room or in an appropriately equipped, dedicated endoscopy suite, ideally under the care of a pediatric anesthesiologist. Securing the endotracheal tube (ETT) and lubrication of the scope present in the oral pharynx is important to reduce the risk of accidental dislodging. Positioning of the tube is also important to meet the needs of the surgeon. With the patient in the left lateral decubitus position, securing the tube to the left side provides additional stability as the tube lies close to the table.

Anesthesia

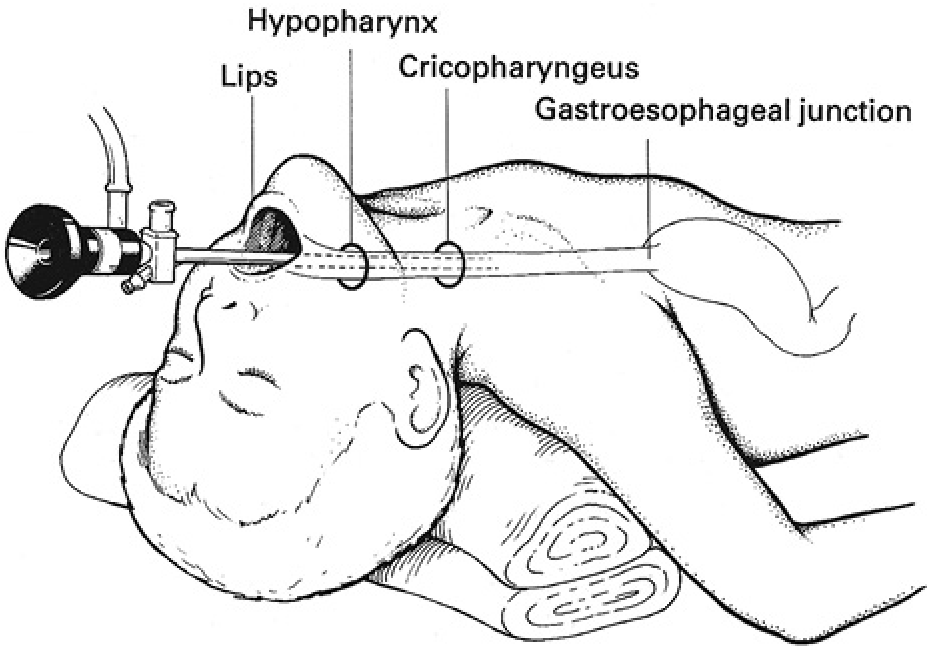
Moderate sedation or general anesthesia can be used. In cases of foreign body removal or in young children where a large endoscope can compress the soft walls of the child's trachea, general anesthesia with endotracheal intubation provides superior airway protection.

OPERATIONS

The following endoscopic techniques are commonly used in pediatrics.

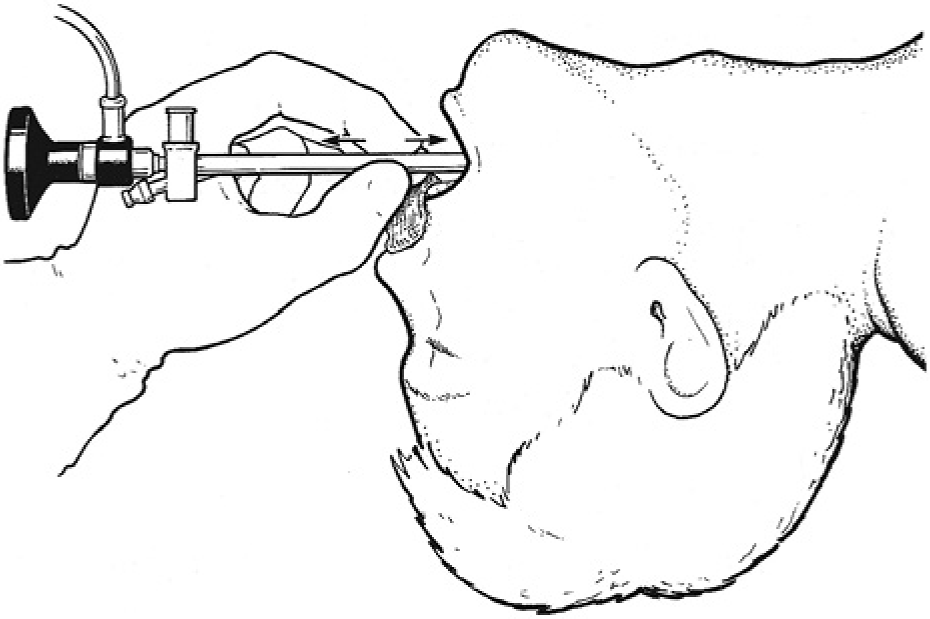
Rigid esophagoscopy

For this procedure, the child is anesthetized and endotracheally intubated (**Figure 6.3**). The ETT has been omitted from the figure for clarity. Supporting towels are placed under the shoulders to maintain the head in full extension. The oropharynx is suctioned. Although the esophagoscope may be passed into the cervical esophagus by direct visualization, this maneuver is facilitated by lifting the tongue and epiglottis with a laryngoscope and directly visualizing the entry into the esophagus. It is critically important that the oroesophageal axis is straight (sword-swallower's position) during rigid esophagoscopy. The teeth, if present, are protected by a gauze pad or plastic guard.



6.3

The esophagoscope is grasped with the supporting hand much as one would grasp a pencil, while the remaining fingers of that hand rest against the maxilla or upper teeth (**Figure 6.4**). The esophagoscope is then slowly advanced, cautiously feeling for resistance. If any difficulty is encountered in negotiating the lumen of the esophagus, a small soft catheter may be used as a lumen finder. Alternatively, gentle insufflation of the esophagus—either with a blood pressure bulb attached to the side of the scope or with CO_2 on low flow—can be used to gently advance the scope. The narrowest points along the way are the cricopharyngeus and the gastroesophageal junction. Once the stomach has been entered, the esophagoscope is slowly withdrawn. It is during the removal of the esophagoscope that the best examination of both the stomach and the esophagus can be obtained.



6.4

Flexible esophagoscopy

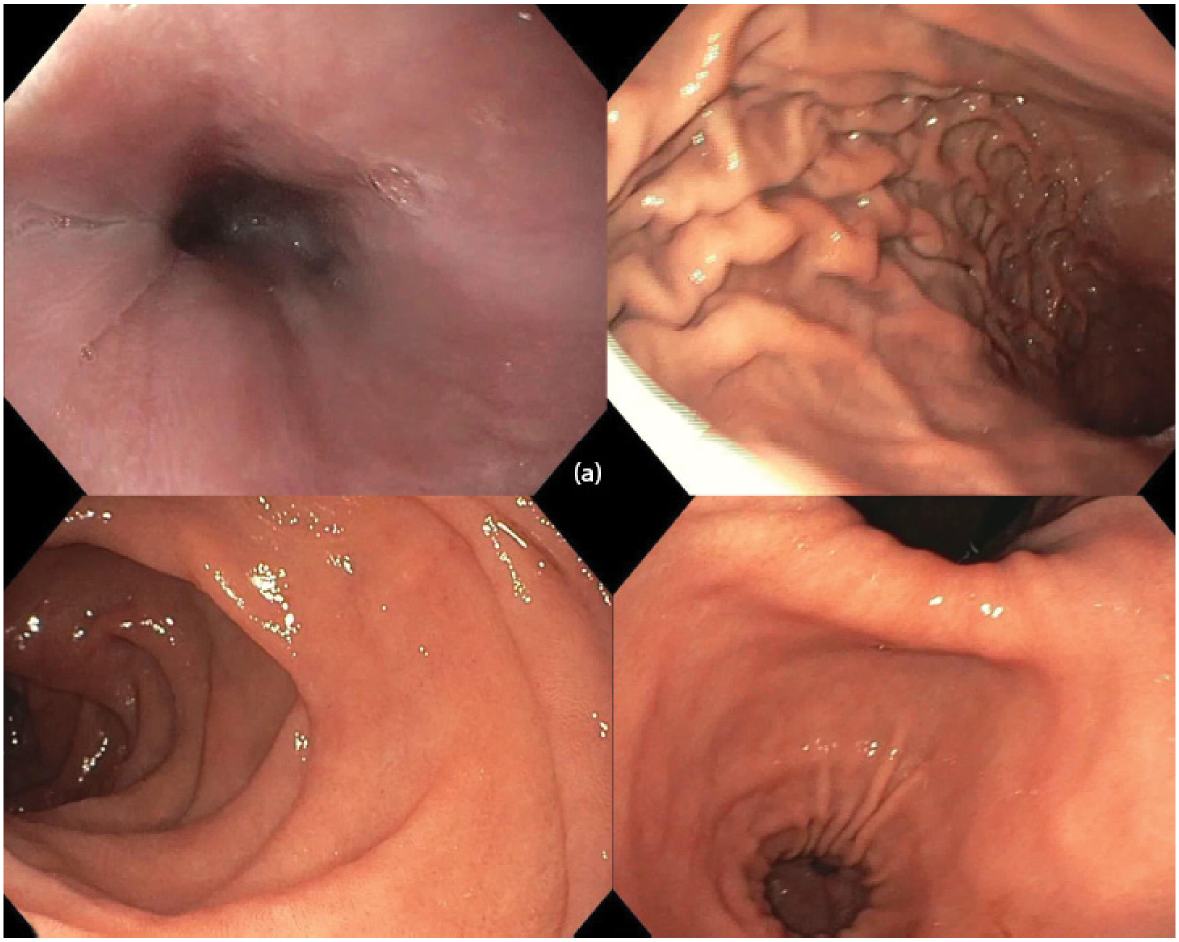
Although conscious sedation can be used in children undergoing flexible esophagoscopy, general anesthesia with endotracheal control of the airway is preferred, especially if procedures such as foreign body removal or dilation are to be performed. The patient can be supine or positioned laterally with the left side down. The endoscope is introduced by directly visualizing the oropharynx and the entry into the esophagus through the cricopharyngeus. A jaw thrust or elevation of the trachea/larynx by directly grasping and elevating it up allows passage of the scope. Advantages of flexible endoscopy include the following:

- Maneuverability allows easier visualization of the entire lining of the esophagus.
- Magnification improves examination of mucosal lesions such as reflux esophagitis or Barrett's esophagus.

- Insufflation of air or carbon dioxide with precise control in order to distend the esophageal lumen is made easier.

It must be emphasized that the same dangers found in using rigid instruments are present during flexible esophagoscopy. The esophagoscope should never be forcibly or blindly advanced. The esophagus may be injured just as easily during manipulations with flexible instruments.

Air insufflation, although invaluable in visualizing the esophagus, must be controlled, especially in infants and small children. Gaseous distension of the abdomen can compromise ventilation. The lowest adjustment possible of the air insufflation rate is used, and the abdomen is left uncovered so that it may be continuously inspected. Carbon dioxide gas may be beneficial compared to air for many endoscopic procedures. After the stomach is entered, insufflation is used to allow a panoramic view (**Figure 6.5**). Excessive fluid is suctioned to allow complete visualization of the mucosa. A gastrostomy, if present, must be occluded to allow the stomach to fill. Orientation and endoscopic maneuvers are similar in the lateral and supine positions. Once the stomach is filled with air, the endoscope can be retroflexed, and the gastroesophageal junction visualized. The endoscope is straightened, and the remainder of the stomach is visualized. With the pylorus in view, the endoscope is advanced. The best visualization of the duodenal bulb and pyloric channel are generally obtained as the scope is first advanced forward with the scope making a large gentle loop along the greater curve (long scope position). As the scope is being withdrawn from the duodenum, this gastric loop reduces first without scope tip movement (short scope position). Once the scope tip begins to withdraw, it generally falls back into the stomach quickly without permitting a proper evaluation of the pylorus and bulb. In a patient with a pronounced angulation at the incisura (the “J-shaped” stomach), a considerable length of the endoscope may need to be advanced before the tip will enter the pylorus. Changing the patient’s position from supine to lateral or from lateral to supine may also be helpful. The ampulla of Vater is recognized by the drainage of bile, but direct visualization of the major papilla is not always possible with a forward-viewing endoscope. Proper evaluation of the lesser curve may require retroflexion in the distal stomach. Insufflated air should be evacuated as the scope is withdrawn.



6.5a–d Endoscopic findings during EGD: (a) Gastroesophageal junction, (b) gastric body, (c) duodenum, (d) partial retroflex view of the lesser curve and the pylorus.

Further evaluation of the small bowel as well as surrounding foregut structures can be performed with double balloon enteroscopy and endoscopic ultrasound.

Grading of esophagitis

The severity of esophagitis can be roughly graded on the basis of the visual appearance alone. Mild esophagitis is indicated by abnormal erythema and friability of the mucosa; moderate esophagitis describes linear erosions and superficial ulcers; and severe esophagitis refers to confluent erosions, deep ulcers, and diffusely hemorrhagic mucosa. The Los Angeles Classification, used for the grading of erosive esophagitis by the number of "mucosal breaks," has proven inter- and intra-user reliability and provides a standardized approach to mucosal evaluation (Table 6.2). Biopsies through the endoscope help diagnose and guide management of such pathologic entities as gastroesophageal reflux, eosinophilic esophagitis, and Crohn's disease.

Table 6.2 Los Angeles Classification for endoscopic grading of esophagitis

Grade A	≥1 mucosal break <5 mm in length that does not extend between tops of two mucosal folds
Grade B	≥1 mucosal break ≥5 mm in length that does not extend between tops of two mucosal folds
Grade C	≥1 mucosal break that is continuous between the tops of 2+ mucosal folds, but involves <75% of esophageal circumference
Grade D	≥1 mucosal break that involves ≥75% of esophageal circumference

Esophageal foreign body removal

COINS

The most common esophageal foreign bodies in children are coins. Other common rigid objects include toy parts and hair beads. Although some advocate fluoroscopically guided balloon extraction of coins as a safe and cost-effective alternative to endoscopic removal, the authors have not adopted this technique. The controlled environment of the operating room is preferred, where adjunctive equipment is readily available to provide airway control. In addition, when the foreign body is removed endoscopically, the esophagus can be directly assessed for damage or other pathology that may have contributed to failure of the foreign body to pass. The most important technical consideration in foreign body removal is the availability of the proper grasping forceps. A "coin forceps" with small teeth at its distal end, allowing it to grasp the edge of a coin or small solid object firmly, is essential (see **Figure 6.1**).

FOOD BOLUS

Removal of an irregular object, such as impacted food material, may be facilitated by the passage of a balloon catheter beyond the material or with the use of a specialized retrieval device (e.g. nets, basket), which is then withdrawn under direct vision while the airway is controlled by endotracheal intubation. Other techniques include the use of suction catheters to pull a soft food bolus back or a “push” technique where the object is gently pushed into the stomach. Food impactions in previously healthy children without known esophageal disease should prompt consideration of eosinophilic esophagitis.

BUTTON BATTERY

Suspected ingestion of a button battery requires emergent evaluation and endoscopic intervention, as these objects can rapidly lead to esophageal damage. It is important to follow recommendations from the National Capital Poison Center which can be found at: https://www.poison.org/battery/guideline. Endoscopic removal should be performed for any button battery in the esophagus. In cases where the battery has passed into the stomach, management algorithms guide therapy. Catastrophic complications including erosion into major vascular structures and/or the airway can occur, and specialized care at pediatric surgery referral centers may be required. Recent evidence supports the immediate administration of honey and/or sucralfate at the time of diagnosis.

MAGNETS

Small, high powered rare-earth magnets are widely available in toys and other common household objects. The incidence of pediatric magnet ingestions has risen dramatically since the early 2000s. When more than one magnet is ingested, the child is at risk for serious complications including bowel obstruction, bowel necrosis and perforation, and fistula formation. Early endoscopic intervention does have some therapeutic role in magnet ingestions, however direct removal via laparoscopy or laparotomy may be necessary.

POSTOPERATIVE CARE

The endoscopic procedure itself may be performed on an outpatient basis. Concurrent or underlying disease may necessitate hospitalization. A chest radiograph is obtained whenever significant manipulation of the esophagus has occurred, such as during a dilatation procedure. The finding of mediastinal air or pneumothorax mandates esophagography with a water-soluble contrast medium to assess the degree of injury. Minor, self-contained perforations may be treated conservatively with antibiotics and hospitalization and possibly pleural drainage. Large perforations with significant pleural or mediastinal communication should be primarily repaired and drained.

ENDOSCOPIC RETROGRADE CHOLANGIOPANCREATOGRAPHY

PRINCIPLES AND JUSTIFICATION

Endoscopic retrograde cholangiopancreatography (ERCP) has a clear role in the diagnosis and management of pancreaticobiliary pathology in the pediatric population, but data on the use of ERCP in pediatric populations are limited due to a much lower incidence of disease than in the adult population. Magnetic resonance cholangiopancreatography (MRCP) provides a non-invasive means to obtain diagnostic information, though it lacks therapeutic potential. In children, the need for anesthetic evaluation/monitoring for conscious sedation during MRI must be considered. As in adult populations, however, ERCP should rarely be used for pure diagnostic purposes and a pre-procedure MRCP to confirm a disease warranting endoscopic indication is generally preferred.

Indications

Over the last decade, the use of therapeutic ERCP has continued to grow while diagnostic ERCP has declined. Diagnostic ERCP has been shown to be useful in the evaluation and operative planning for certain conditions including choledochal cysts, pancreaticobiliary maljunction, and biliary atresia. Therapeutic ERCP has been shown to successfully manage numerous causes of biliary obstruction in children. Sphincterotomy, stone extraction, stent placement, and biliary dilation can be used in appropriately selected patients. Conditions including choledocholithiasis, biliary strictures, and bile leak following trauma, cholecystectomy, or hepatic surgery can be appropriately managed with ERCP. Ductal stone clearance in pediatric choledocholithiasis has been consistently shown to have a success rate greater than 90%.

Management of pancreatic ductal pathology is principally similar to biliary management, with sphincterotomy and ductal stenting/dilation effective for numerous pediatric conditions. Complications of pancreatic pathology including pseudocyst, ductal leak, fistula, and stricture have been managed successfully in children.

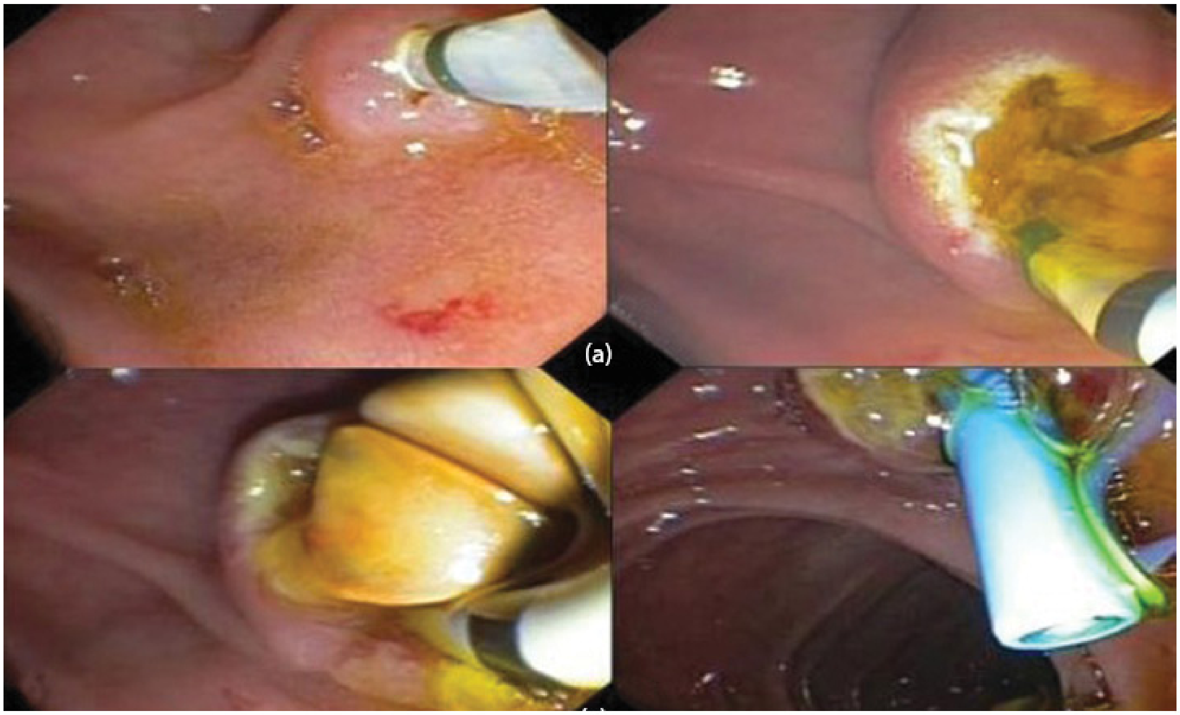
Most commonly, adult duodenoscopes are utilized to perform ERCP in children, but their large outer diameter may preclude their use in smaller patients. The availability of pediatric-sized duodenoscope is limited, even at major academic centers, and this unavailability has implications on the ability to perform ERCP on the smallest of patients.

PROCEDURE

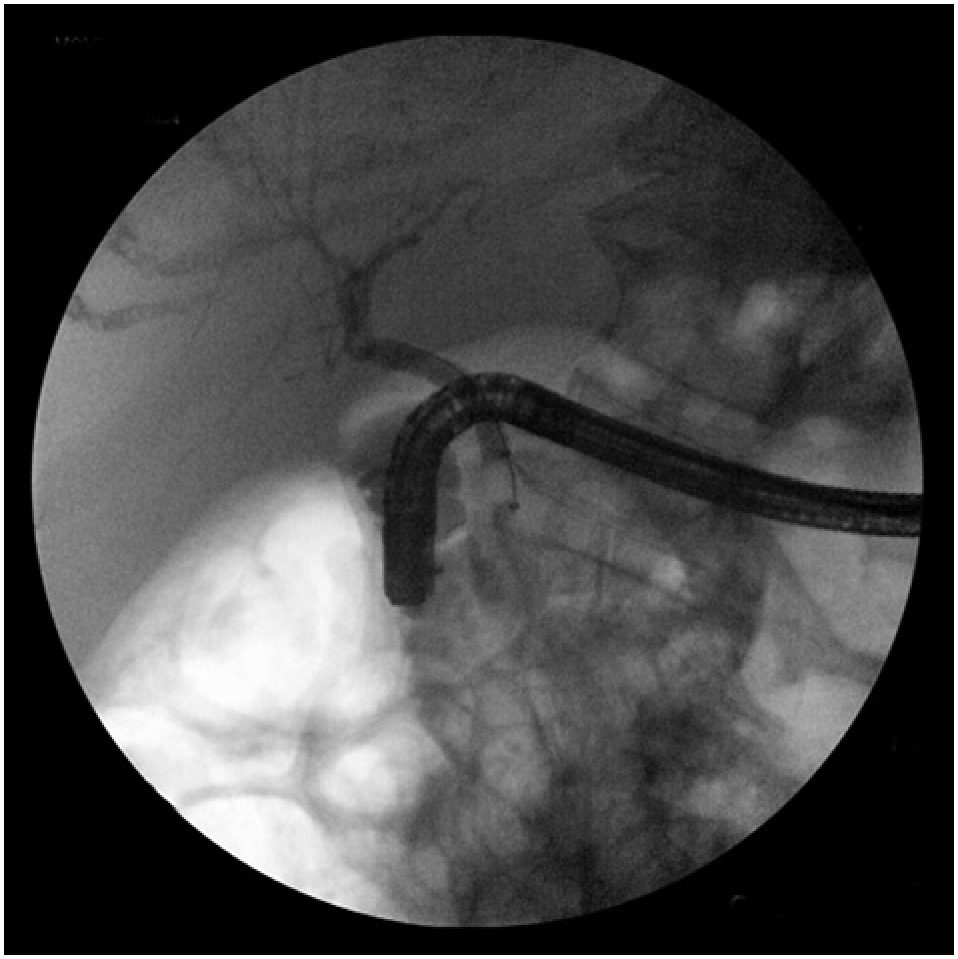
The principles of perioperative care for pediatric patients during ERCP are largely similar to other forms of endoscopy. Moderate sedation or general anesthesia can be used. The use of an oversized duodenoscope can compress the soft walls of a child’s trachea; providers choosing moderate sedation must be cognizant of this risk. Positioning of the child in the modified swimmer’s position facilitates easier scope passage and better access to the sphincter of Oddi. A pre-procedure scout fluoroscopic image should be taken before scope introduction.

ERCP is performed with a side-viewing endoscope. For children greater than 15 kg, adult duodenoscopes with diameters of 10.6 mm or greater are typically used. Pediatric duodenoscopes, with diameters less than 7.5 mm, are available (albeit limited). The side-viewing duodenoscope must be introduced into the esophagus with great caution. Because the scope is advanced with a limited view, any encountered resistance should prompt the use of a forward-viewing endoscope to evaluate for anatomic causes.

After passing the pylorus, the major papilla is typically visualized on the medial wall of the duodenum (**Figure 6.6 a–d**). Similar to EGD procedures, the endoscope traverses the pylorus in a long position and is then transitioned to a short scope position prior to procedural intervention. Once identified, a multi-lumen catheter or sphincterotome is advanced into the lumen of the papilla in the anticipated location of the desired orifice (biliary or pancreatic). A careful visual inspection, anatomical knowledge, and selective positioning of the scope allow for proper cannulation of the biliary or pancreatic duct. Following deep selective cannulation of the desired duct, a guidewire is advanced under fluoroscopic guidance. Contrast injection is utilized as needed to define anatomy and pathologic alterations warranting intervention (**Figure 6.7**). Pancreatic duct injection, whether intentional (to define pancreatic ductal anatomy) or unintentional (as a consequence of an attempted biliary duct cannulation), can result in post-ERCP pancreatitis. Unintended injection should be avoided by selective bile duct cannulation. Intentional injection should occur under low pressure and with as low a volume of contrast as possible. Over the guidewire, specific devices including a sphincterotome, extraction balloons or baskets, stents, or balloon dilators can be advanced to accomplish the therapeutic aims of the procedure. A completion fluoroscope image should be taken to confirm contrast location, to document bile duct emptying, and to confirm stent position (if placed).



6.6a–d Endoscopic view during ERCP: (a) Cannulation of the major papilla, (b) sphincterotomy, (c) extraction of biliary calculi, (d) stent placement.



6.7 Fluoroscopic image during ERCP; contrast injection of the bile duct confirms a healed cystic duct stump leak after cholecystectomy.

POSTOPERATIVE CARE

Complications from ERCP, although rare, do occur more frequently than from upper endoscopy alone. Bleeding and perforation, which occur in less than 1% of cases, are associated with sphincterotomy and can often be managed endoscopically. Post-ERCP cholangitis is associated with incomplete biliary drainage. Data support the use of prophylactic antibiotics only in cases where an obstructed (but not infected) bile duct is expected to remain obstructed at the conclusion of the procedure and where wire or catheter passage may inoculate the obstructed bile collection and result in infection. Post-ERCP pancreatitis (PEP) is defined as a serum amylase level greater than three times the upper limit of normal with accompanying abdominal pain. In studies of children, 3–11% of ERCPs are complicated by PEP. PEP management is identical to the management of acute pancreatitis of other causes; most cases can be managed with bowel rest and IV hydration. Prophylactic measures to prevent PEP, such as rectal indomethacin use or prophylactic pancreatic stent placement, have not been studied in children.

VIDEO

OPS_8e_video_6-1 ERCP

FURTHER READING

Anfang R, Jatana K, Linn R, et al. pH-Neutralizing esophageal irrigations as a novel mitigation strategy for button battery injury. *Laryngoscope* 2019; 129(1): 49–57.

Baskin D, Urganci N, Abbosoglu L, et al. A standardized protocol for the acute management of corrosive ingestion in children. *Pediatr Surg Int* 2004; 20: 824–8.

Gans SL. Principles of optics and illumination. In: Gans SL (ed.). *Pediatric Endoscopy*. New York: Grune and Stratton; 1983: pp. 1–8.

Lundell L, Dent J, Bennett J. Endoscopic assessment of oesophagitis: Clinical and functional correlates and further validation of the Los Angeles classification. *Gut* 1999; 45: 172–80.

National Capital Poison Center. Battery Ingestion Triage and Treatment Guideline. https://www.poison.org/battery/guideline (Accessed 16 February 2020).

Rosen J, Lane R, Martinez J, et al. Success and safety of endoscopic retrograde cholangiopancreatography in children. *Pediatr Surg* 2017; 52: 1148–51.

7

Colonoscopy

IAN D. SUGARMAN AND JONATHAN R. SUTCLIFFE

HISTORY

The first colonoscopies were performed in 1969 by surgeons William I Wolff and Hiromi Shinya working at the Beth Israel Medical Center, New York City in collaboration with the Olympus Optical Company. Three months after the first colonoscopy, the first colonoscopic snare was performed and this led the way to a major change in the screening and treatment of colorectal cancers which continues today.

Paediatric colonoscopy was first reported in the 1970s and has now become the standard investigation for lower gastrointestinal (GI) symptoms. Over time, the colonoscopes have become more sophisticated to allow better training and maneuverability in the colon.

PRINCIPLES AND JUSTIFICATION

Indications

The indications for colonoscopy can be divided into diagnostic, screening, surveillance, and therapeutic. In diagnostic cases, the commonest indication is to investigate symptoms suggestive of inflammatory bowel disease (IBD). These include failure to thrive or weight loss, chronic diarrhea, anemia, bleeding, mucous per rectum (PR), or when there is radiologic abnormality (e.g. narrowed terminal ileal abnormality on ultrasound, small bowel follow-through, or magnetic resonance imaging (MRI)). The second commonest indication is the investigation of painless rectal bleeding to exclude the presence of rectal/colonic polyps with the next commonest being screening and surveillance of children with polyposis syndromes until a plan for surgical intervention is required. Rarely, the indications include the investigation of tenesmus in order to exclude a solitary rectal ulcer and non-specific abdominal pain (with a very low pick-up rate).

The current treatment of IBD has, as one of its main aims, mucosal healing. In contrast to radiological examination, colonoscopy allows visualization of the mucosa and histological assessment. When indicated, it is feasible to perform both colonoscopy to the terminal ileum and gastroscopy to the duodenum at a single examination. A large proportion of the GI tract is thus rendered accessible to inspection, biopsy, or instrumentation in one procedure. Performing an esophagogastroduodenoscopy (EGD) at the same sitting as colonoscopy is particularly useful for children with potential IBD. In these children, evidence of Crohn's disease may exist in the upper GI tract before it appears in the lower GI tract.

Contraindications

There are few contraindications to colonoscopy in experienced hands with appropriate instrumentation. Examination is unlikely to be helpful in constipation and, as stated above, the diagnostic yield is extremely low in abdominal pain unaccompanied by features to suggest systemic illness. There is a risk of translocation leading to septicemia in marasmic, immunodepressed, or immunosuppressed subjects, who should receive appropriate antibiotics. There is a risk of bacterial peritonitis when colonoscopy is performed in the presence of ascites.

PREOPERATIVE

Bowel preparation

A variety of bowel preparation regimens for children are now available that will produce a clean colon. In our institution we use Picolax™ in doses dependent on the child's age, or Senokot® in children aged <1 year (Table 7.1).

Table 7.1 **Suggested bowel preparation**

Age (years)	Medication	1st dose (24 hours pre-op)	2nd dose (18 hours pre-op)
<1	Senna (e.g. Senokot)	5–10 mL	5–10 mL
<2	Picolax	1/4 sachet	1/4 sachet
2–5	Picolax	1/2 sachet	1/2 sachet
6–10	Picolax	1 sachet	1/2 sachet
>10	Picolax	1 sachet	1 sachet

Sedation versus anesthesia

Premedication may be useful for apprehensive children, for whom reassurance and explanation are often ineffective. Colonoscopic examination may, however, be uncomfortable or painful during the procedure as the instrument stretches the sigmoid colon mesentery or the visceral peritoneum. General anesthesia rather than sedation is therefore recommended for most children.

Choice of instrument

There is an increasing array of colonoscopes available in terms of size, length, variable stiffness, definition and magnification, and narrow band imaging, however a summary of what is used at our institution is given in Table 7.2, with their characteristics.

Table 7.2 **Lower gastrointestinal endoscopes**

Name	Make	Age of patient (years)	Weight (kg)	Outside diameter (mm)	Working channel diameter (mm)	Length (mm)	Variable stiffness	HD[a]
EG450PE	Fujinon	≤2	<10	8.2	2.2	1100	No	No
PCF240I	Olympus	≤8	≥10	10.3	3.2	1330	No	No
CF240AL	Olympus	≥6	≥17	12.2	3.2	1680	Yes	No
CFQ260AL	Olympus	≥8	≥22	13.2	3.7	1680	Yes	No
2018 release								
GIFH290	Olympus	≤2	<10	8.9	2.8	1030	No	Yes
PCFH290TLI	Olympus	≥2	≥10	10.5	3.2	1330	Yes	Yes
PCFH290DL	Olympus	≥6	≥17	11.8	3.2	1680	Yes	Yes
PCFH290ZL	Olympus	≥8	≥22	11.8	3.2	1680	Yes and zoom	Yes

[a] High definition.

Is radiographic control needed?

Most examinations do not require radiographic screening control and the majority of colonoscopists never use it. It has been shown that magnetic endoscopic imaging (which involves either passing a magnetic probe through the biopsy channel, or in newer colonoscopies, the scope itself has metal markers which can be seen with appropriate screening monitors. This is a useful tool for training and dealing with the more difficult loops that may occur as it shows the progress of the colonoscope, reveals when loops are developing, and helps the operator in learning how to either prevent or remove a loop for quicker progress along the colon.

OPERATIONS

Colonoscopy

POSITION OF PATIENT

Infants are usually examined supine while in older patients the left lateral position is the norm, and it is often possible to complete the examination without a position change. If there are mechanical difficulties at any stage of the procedure, a change in position may alter the configuration of the bowel and facilitate examination. Such movement of a larger child under general anesthesia may require a number of people to be present. Changing to the right lateral position will make the splenic flexure less acute and can also help to drain fluid from the descending colon and facilitate air distension within it if the view is poor. Soft padding is used to protect pressure areas at all times.

INITIAL STEPS: INSERTION AND PASSAGE THROUGH THE RECTOSIGMOID

An important component of the examination is palpation to exclude abdominal masses in the right lower quadrant (suggestive of terminal ileitis in Crohn's disease) and perineal inspection. The presence of tags, deep fissures, and induration makes the diagnosis of Crohn's disease more likely and this information may help differentiation from ulcerative colitis. Prior to intubation of the anus, a rectal examination should be performed to exclude a distal polyp. Proctoscopy not only allows visualization of the distal rectum but also allows liquid stool to be drained from within the rectum which in turn improves examination of the rectal ampulla.

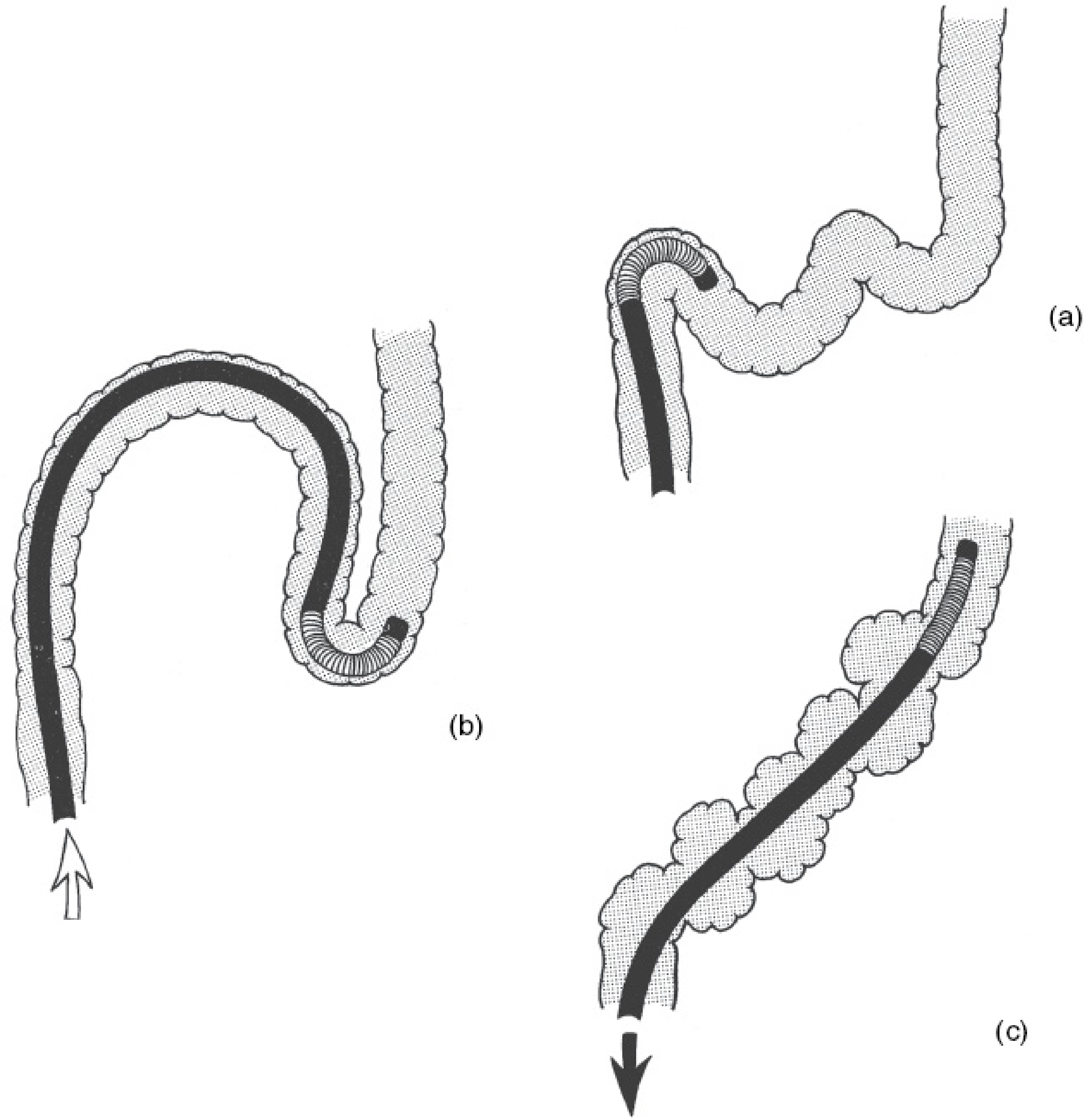
The tip of the colonoscope and the perianal region are lubricated with jelly. On insertion, initially there may be no view because the tip is against the wall of the rectum. The instrument must be withdrawn slightly, and air insufflated before a view is obtained, the tip then being angled, and the instrument shaft rotated as necessary to follow along the lumen of the rectosigmoid. In passing the many bends of the rectosigmoid, the object is to avoid distending or stretching the bowel so as to keep it short and pass almost straight to the descending colon. This is easier to suggest than to achieve but is made more likely by observing the following points.

- As little air as possible should be insufflated to see; excess air should be aspirated from time to time.
- The bowel lumen should be followed accurately.

- If the view is lost, even for a few seconds, the control knobs must be released and the colonoscope withdrawn a short distance—the lumen will automatically reappear.
- A number of bends will be encountered. To navigate around a bend, first identify a fold of mucosa forming a 180-degree arch at the bend. Rotate the scope so that this bend is at the 12 o'clock position, deflect the tip of the scope up to the 12 o'clock position, and advance.
- Blind pushing should be avoided, but on acute bends this may be necessary for a few seconds providing the mucosa continues to move and the general direction is known.
- If the tip will not angle round a bend, an attempt should be made to "corkscrew" the instrument by pulling the shaft back straight and twisting it, at the same time, one way or the other to keep the tip of the scope in the centre of the lumen.
- If large loops have been made, the colonoscope should be pulled back repeatedly after passing a bend and before starting each inward push. A straight colonoscope and a shortened colon will result.
- The instrument shaft should be held in the fingertips as far as possible—gripping in a clenched fist causes clumsiness.

SIGMOID N-LOOP: HOOK AND TWIST MANEUVER

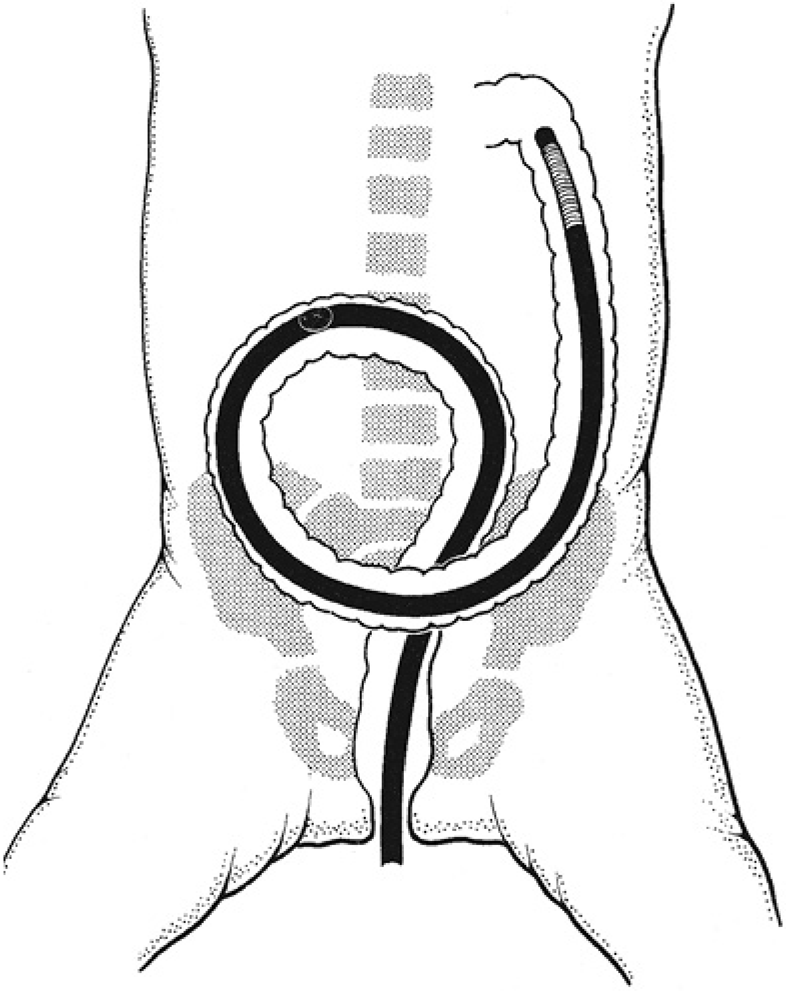
The commonest situation on reaching the junction of the sigmoid and descending colon, in spite of all care, is for there to be an N-loop forming an acute tip angle which makes direct passage difficult or impossible (**Figure 7.1a**). If the tip can be passed a short way around the bend, looking in to the retroperitoneal part of the descending colon, it can be held there without consciously hooking while the instrument is withdrawn 10–40 cm to reduce and straighten out the loop (**Figure 7.1b**). Putting a clockwise twisting force or torque on to the shaft of the colonoscope while it is withdrawn will help to straighten out this loop and keep the tip in the descending colon (**Figure 7.1c**).



7.1a–c

SIGMOID “ALPHA” LOOP

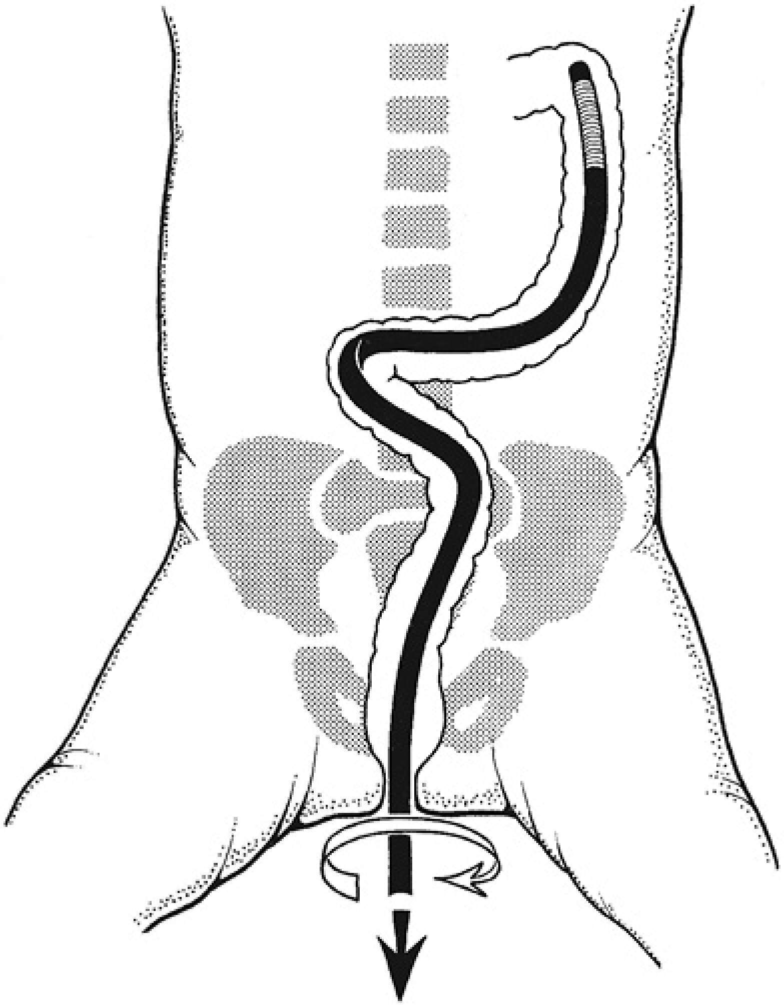
Often, if there is a redundant colon, a loop is obviously forming but the tip runs in easily without discomfort to the patient. This suggests that a spiral “alpha” loop is forming (**Figure 7.2**). The correct thing to do is to continue pushing (in as far as is comfortable for the patient if awake), at least to the proximal descending colon and preferably to the splenic flexure. Once around the splenic flexure, the instrument can be "hooked" over the flexure while attempting to withdraw and straighten it out to remove the loop as described below.



7.2

STRAIGHTENING OUT LOOPS

Having reached the transverse colon, the sigmoid colon loop should be removed since loops create friction in the control wires and stress the instrument just as much as they stress the patient. To remove a loop, ensure that the tip of the scope is held in the centre of the lumen at all times. The instrument shaft should be withdrawn until the tip begins to slide past the mucosa or resistance to withdrawal is felt. While pulling back, the application of twist, usually in a clockwise direction, will stop the tip slipping back excessively and facilitate the straightening of the instrument (**Figure 7.3**). Once 1:1 movement is obtained (i.e. for each centimeter of movement of the scope by the operator there is a commensurate centimeter of movement at the tip of the scope), the scope can be readvanced. Leaving the same direction of torque on the scope as it is advanced is likely to allow reinsertion without reforming a loop. It is sometimes possible to ask an assistant to apply external pressure to the apex of a loop (often higher than might be expected) which in turn reduces the likelihood of a loop progressing.

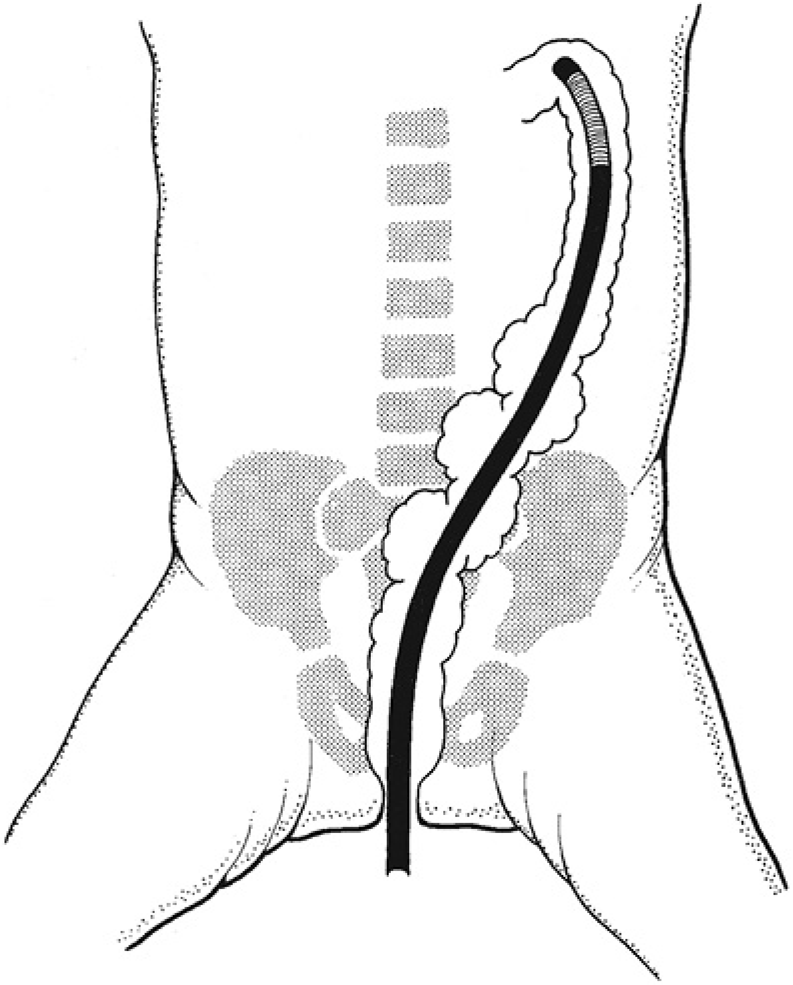


7.3

In the young child, it is very likely that the colon will prove to be hypermobile, without conventional fixation of the descending colon and splenic flexure. In the 20–30% of patients who have a mobile colon, unpredictable and sometimes uncontrollable loops may form.

SPLENIC FLEXURE: KEEPING THE SIGMOID COLON STRAIGHT

With the colonoscope straightened in the proximal descending colon or splenic flexure, some care may be needed to prevent the sigmoid loop reforming. Continued clockwise (or sometimes anticlockwise) twist on the shaft during reinsertion is often enough to keep it straight (**Figure 7.4**). It is at this point that the variable stiffness colonoscopes come into their own. Once the loop is removed, by increasing the stiffness of the scope, the chance of the loop recurring is diminished. Shaft insertion without tip movement or losing the 1:1 relationship between shaft and tip indicates looping. The instrument is immediately pulled back again, and the assistant pushes into the left iliac fossa to reduce the tendency for the sigmoid loop to rise up from the pelvis.



7.4

In the splenic flexure, this tendency to re-loop in the sigmoid colon results because of the resistance of the hooked instrument tip in the splenic flexure. A combination of the following small corrective measures will usually overcome this:

- The instrument shaft should be pulled back straight.
- If possible, the scope should be stiffened.
- Hand pressure should be applied by the assistant over the left iliac fossa.
- The instrument shaft should be twisted clockwise.
- If necessary, the instrument should be re-aimed toward the lumen, avoiding over-angulation.

- Clockwise torque should be applied as the scope is slowly advanced.

Sometimes it is easier to reposition the child in the right lateral position to cause the splenic flexure to drop down and flatten out.

REDUNDANT TRANSVERSE COLON

The transverse colon may sometimes be pushed down by the instrument into a deep loop, which makes it difficult and painful to reach the hepatic flexure. Once again, the correct maneuver is to withdraw the instrument to shorten this loop. If necessary, withdrawal may need to be repeated several times, the instrument advancing a few centimeters on each withdrawal (“paradoxical movement”) until the loop is straightened. Keeping the colon deflated also helps to shorten the hepatic flexure region, making it easier both to reach and to pass. In addition, an assistant pushing the transverse colon upwards and straightening this loop out is often helpful.

Difficulty in the transverse colon may also be due to recurrent looping in the sigmoid colon, and the best corrective measures are abdominal pressure in the left iliac fossa and gentle clockwise twisting during reinsertion.

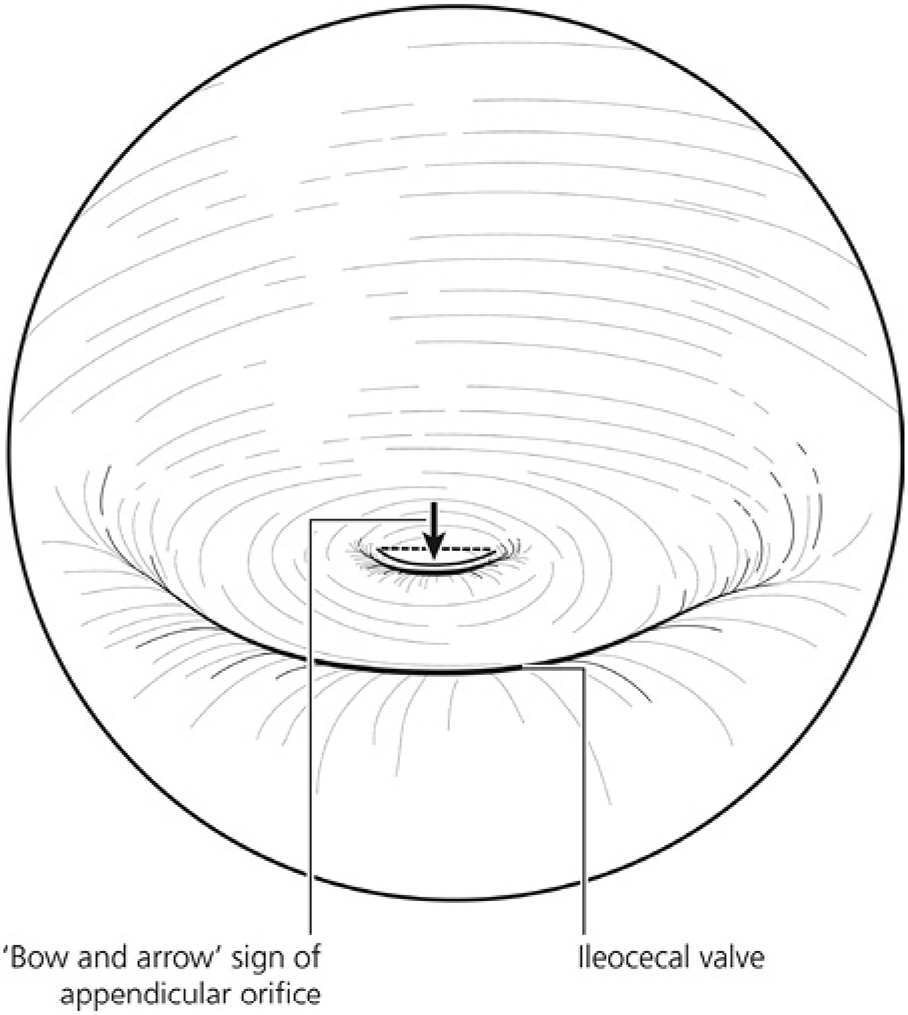
PASSING THE HEPATIC FLEXURE

Do not mistake the hepatic flexure for the cecum; the presence of the liver often produces a blue discoloration visible through the wall of the colon, but if doubt remains, briefly increasing the intensity of the scope light will allow the tip of the scope to be located in the right upper quadrant.

Having reached and deflated the hepatic flexure, and angled acutely around it into the ascending colon, the transverse loop may remain and make it difficult to pass the rest of the instrument into the ascending colon. By once again withdrawing the colonoscope and straightening out this loop, it becomes easier to pass. Deflating the ascending colon by aspiration and simultaneously steering carefully to avoid haustral folds will often cause the colonoscope to descend spontaneously toward the cecum.

REACHING THE CECUM

Reaching the cecal pole can be facilitated by sucking out the gas and deflation of the colon, application of abdominal pressure in the left lower quadrant, and clockwise twist on the straightened instrument. The scope can often then be gently advanced; aggressive pushing usually only results in looping. The colonoscope is seen to have reached the cecum when the bulge of the ileocecal valve is seen or, 2–5 cm beyond it, the appendicular orifice is identified (**Figure 7.5**). The ileocecal valve is the largest fold lying close to the cecal pole and can often be identified by following the arrow marked in **Figure 7.5**, in the "bow and arrow" sign of the appendicular orifice. Brilliant transillumination in the right iliac fossa is usually apparent at this point.



7.5

The depth of insertion of the straightened instrument is variable according to the age of the patient: 70–80 cm in a teenager, down to 25 cm in a small infant. During withdrawal, the splenic flexure or descending colon is found at appropriately shorter distances. During insertion, in mobile colons or if any loops have been formed, these distance rules may not apply, but if the room is darkened, transillumination will show the position of the instrument tip.

To enter the terminal ileum, it is necessary first to identify the bulge of the ileocecal valve, which may bubble or gush on deflation. The instrument tip is then pushed in just proximal to the bulge, angled in toward it, and slowly withdrawn until a "red-out" indicates embedding into the valve region, at which point air is insufflated to attempt to distend the ileum.

Alternatively, the biopsy forceps are advanced through the end of the scope, which is placed such that the ileocecal valve lies adjacent to the 6 o'clock position (and therefore closest to the scope's biopsy forceps channel). The tip of the scope is positioned just beyond the ileocecal valve, the biopsy forceps advanced 1 cm and then the tip angulated down slightly. As the scope is gently withdrawn, the biopsy forceps will often enter the ileocecal valve. If the forceps are then advanced by 1–2 cm, the scope itself can then be advanced over the forceps thus entering the ileum.

Ileal mucosa is characteristically granular or nodulated by lymphoid hyperplasia, in contrast to the shiny surface and vascular pattern of the colon.

In infants under 1 year of age, entry into the ileum may be impossible, either because the orifice is too narrow or because the dimensions of the cecum are too small to allow the instrument to make the necessary right-angle turn.

EXAMINATION

The colon is visualized to some extent during insertion of the instrument, but active examination, biopsy, or polypectomies are normally undertaken during withdrawal because the instrument is then straight and easy to maneuver, the view is better, and the patient is more comfortable. At all stages during the examination, but particularly during withdrawal, it is best for the endoscopist to control the instrument, using a one-handed technique. Very active maneuvering of the controls, with rotation and to-and-fro movements of the shaft, allows a good view to be obtained of nearly all areas, although around acute bends and convoluted folds there may be some blind spots. Ensure representative photographs are taken throughout this phase of the examination.

BIOPSIES

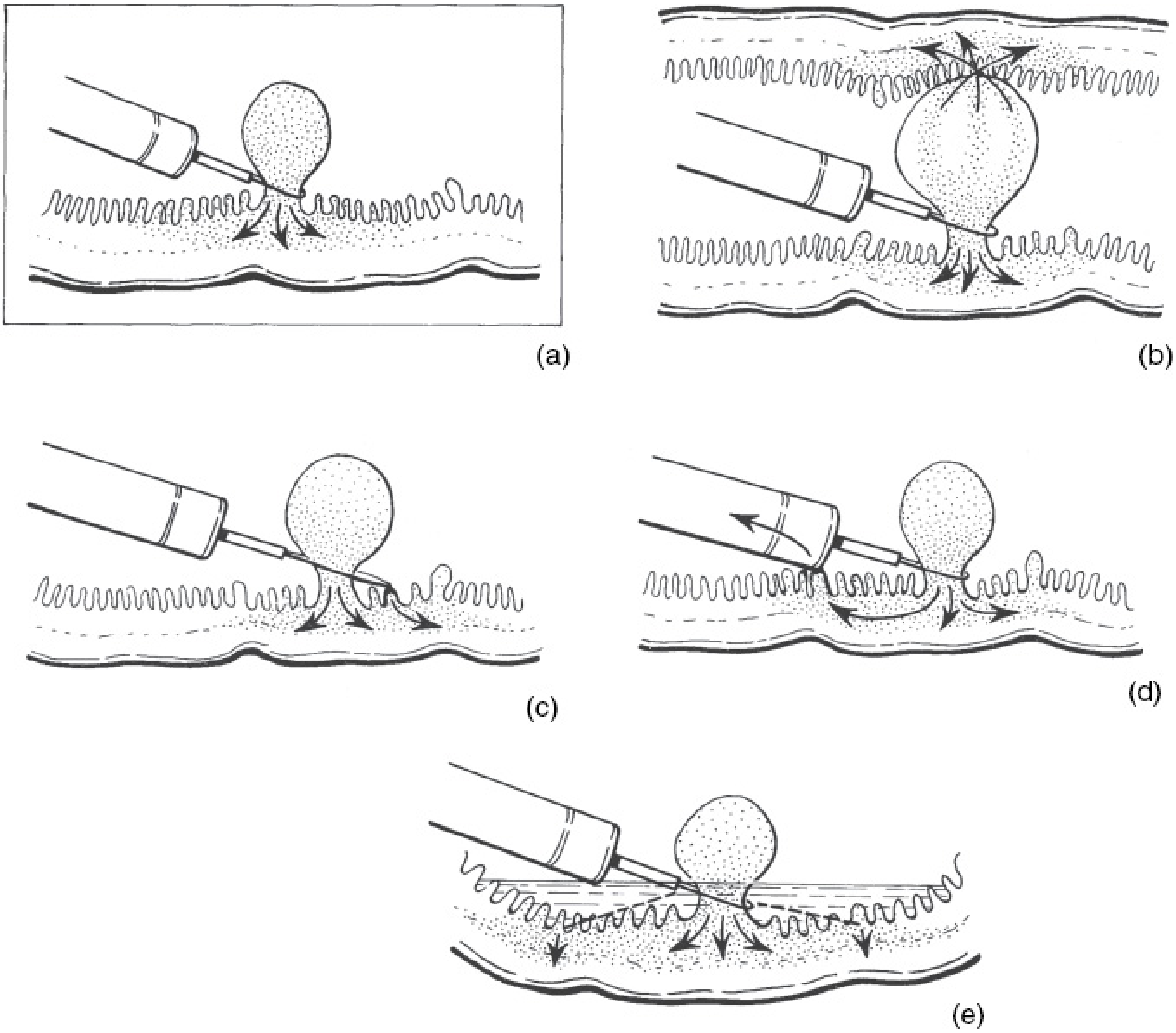
Most colonoscopies are diagnostic and made both on the macroscopic and microscopic findings. It is our practice, at the pathologist's request, to take two specimens from any segment as this helps to assess the histological patchiness of inflammation and may help in distinguishing between Crohn's disease and ulcerative colitis. Our standard practice is to take biopsies from the terminal ileum, cecum, ascending colon, transverse colon, sigmoid, and rectum. If localized pathology is seen in a segment, abnormal and adjacent macroscopically normal colon should be biopsied.

COLONOSCOPIC POLYPECTOMY

The principles of colonoscopic polypectomy are identical to those for proctosigmoidoscopic polypectomy, but it is particularly important that full coagulation of polyp stalk vessels is achieved before transection, as any hemorrhage is difficult to control endoscopically. Most polyps in pediatric practice are hamartomatous, thin-stalked, and easy to coagulate. If a thick stalk (≥1 cm) is to be snared, it may be wise to inject it with epinephrine (adrenaline) (1 mL of 1 : 100 000 solution), using a long Teflon sclerotherapy needle before applying the polypectomy snare. Alternately, a ligating loop can be placed on the proximal stalk before the more distal stalk is divided with diathermy snare.

Endoscopic snare wires are characteristically thick, to guard against cutting too fast, but care should be taken not to apply excessive mechanical pressure before adequate electrocoagulation has occurred, otherwise "cheese-wiring" of an uncoagulated stalk may occur with consequent hemorrhage. A low-power coagulating current (15–25 W) is employed until local whitening or swelling of the stalk indicates adequate coagulation, at which point tight strangulation should result in severance of the head. If bleeding does occur, the stalk remnant can be quickly re-grasped with the loop and strangulated for 15 minutes, after which bleeding will not normally recur.

The correct position of the snare is shown in **Figure 7.6a**. Care must be taken to avoid contact of the polyp surface with the opposite wall leading to burns from dissipation of the current (**Figure 7.6b**). Burns may also result if the active electrode or metal components of the colonoscope tip are in contact with the local tissue (**Figure 7.6c and d**) or if the electrode is in contact with a pool of fluid (**Figure 7.6e**).



7.6a–e

Rarely, sessile polyps are found. These may be removed by endomucosal resection (EMR), but this is an advanced technique and beyond the scope of this chapter.

Other therapeutic maneuvers

The colonoscope can be used to introduce guidewires, tubes, and other devices to any point in the colon. One therapeutic use of the colonoscope is that of insertion of an antegrade colon enema (ACE) button to allow colonic irrigation. The button can be placed in the cecum (ACE) or sigmoid (DACE) and is performed in two stages. First, inserting a tube and then, six weeks later, changing the tube for a button. The first stage is performed exactly as per insertion of a percutaneous endoscopic gastrostomy (PEG) tube and in the cecum is a relatively easy and quick procedure. It is more challenging in the sigmoid as it difficult to keep the sigmoid insufflated compared to the cecum (or stomach). We place a 15 Fr Freka^{TM} tube (Fresenius Kabi, Runcorn, UK) first such that, when changed to a 14 Fr MIC-KEY^{TM} or MINI button^{TM} (Avanos Medical UK, Redhill, Surrey, UK), the button slips in with no resistance.

Electrocoagulation of a telangiectasia or cavernous hemangioma (blue rubber bleb nevus syndrome) is easy through the colonoscope. The use of laser photocoagulation for this purpose has been described but is probably unnecessary, since careful scleropathy of raised lesions, repeated as necessary, gives excellent results.

Strictures, particularly anastomotic strictures after resection of Crohn's disease, can be successfully dilated with transendoscopic balloon dilators, following which, if indicated, absorbable stents can be placed. The use of submucosally injected Indian ink can be useful as a long-lasting marker. Surface irrigation with colorant (1:4 dilution of washable blue fountain pen ink is convenient) can be helpful in demonstrating the smallest lesions in conditions such as familial adenomatous polyposis.

POSTOPERATIVE CARE

In most cases, no special care is needed after colonoscopy, apart from a short period of rest until the after-effects of sedation or anesthesia wear off. Food and drink can be taken immediately. When the patient appears and feels well, normal activities can be resumed, with many examinations being performed on a day-case basis.

Follow-up is probably unnecessary after colonoscopic polypectomy if only one to four juvenile polyps are present in the colon; larger numbers may suggest the possibility of juvenile polyposis, which mandates follow-up because of the association with dysplastic foci. The other commonest group who undergo surveillance is children with familial adenomatous polyposis prior to corrective surgery.

COMPLICATIONS

The main risk is that of perforation. The risk is about 1 in 1000 for a diagnostic colonoscopy, with an increased risk for polypectomies. The presence of severe acute IBD with peritonism or deep ulceration, however, increases the possibility of perforation. Even in a normal colon, if the procedure proves technically difficult for example due to poor "bowel prep" or a difficult-to-manage loop, common sense may suggest abandonment of an examination.

The highest percentage of complications occurs following therapeutic maneuvers such as snare polypectomy, when both perforation and bleeding have been reported. It is recognized that delayed perforation after polypectomy may occur, and this may also be seen after a "hot biopsy" which is why EMR is now the treatment of choice in adult practice.

Infantile colonoscopy is the most difficult technique to accomplish successfully but in children and adolescents cecal intubation is achieved in >90% and ileal intubation in >85%.

OUTCOME

Diagnostic yields from colonoscopy vary depending on symptoms at presentation. Our review of over 400 lower GI endoscopies showed that diagnostic yields for children with any rectal bleeding ranged from almost 85% (PR bleeding, abdominal pain, mucous, and diarrhea) to under 50% (PR bleeding alone), but on our unit review an even lower yield (<30%) was found when the only symptom was non-specific abdominal pain. One or more polyps were found in about 10% of this series.

Both colonoscopic polypectomies and colonoscopic cecostomy tube insertion are safest when performed by experienced personnel. This may be our adult gastroenterology colleagues when EMR for larger polyps is required.

VIDEO

OPS_8e_video_7-1 Colonoscopic ACE

FURTHER READING

Campbell AM, Sugarman I. Does painless rectal bleeding equate to a colonic polyp? *Arch Dis Child* 2017; 102: 1049–51.

Cotton PB, Williams CB. *Practical Gastrointestinal Endoscopy: The Fundamentals*, 6th ed. Hoboken, NJ: Wiley-Blackwell; 2008.

PART III

Thoracic

8

Principles of thoracic surgery

SHAUN M. KUNISAKI AND JAMES D. GEIGER

PRINCIPLES AND JUSTIFICATION

This chapter describes the basic operative approaches to the chest in pediatric thoracic surgery. These approaches include traditional posterolateral thoracotomy, muscle-sparing thoracotomy, axillary thoracotomy, median sternotomy, and, more recently, thoracoscopy. The skilled pediatric surgeon should be comfortable with all of these modes of access. Based on the underlying pathology, a properly chosen incision can provide optimal exposure of the structure of interest. Conversely, the wrong thoracic approach can lead to poor visualization of the operative field with unnecessary intraoperative and postoperative morbidity.

ANESTHESIA

General endotracheal anesthesia is required for most intrathoracic operations. The notable exception to this rule would be a biopsy in a child with tracheal compression from a giant anterior mediastinal mass. In this case, local anesthesia would be advised because of the risk of complete airway loss and cardiovascular collapse secondary to caval compression associated with using muscle relaxants upon induction.

Following the administration of general anesthesia, tracheal intubation can be sufficient for many open and minimally invasive procedures, such as thoracoscopic pulmonary decortication, because of the relatively easy visualization of the pleural space following collapse of the ipsilateral lung with carbon dioxide insufflation. In these situations, having the anesthesiologist ventilate with smaller tidal volumes, lower peak pressures, and high respiratory rates can sometimes further enhance exposure.

However, there are many cases, such as in thoracoscopic pulmonary lobectomy, in which selective intubation to better isolate the lung is preferred. These techniques require an experienced anesthesiologist. Intubation of the contralateral mainstem bronchus with an uncuffed tube is one, relatively easy option for selective intubation in all age groups. However, one drawback is that mainstem intubation does not always give complete lung isolation unless there is a good seal and no overflow ventilation.

Another well-described technique is the bronchial blocker, which is a 5 Fr catheter that is deployed to occlude the ipsilateral mainstem bronchus. Bronchial blockers have a high-volume, low-pressure balloon to isolate the lung but require a least a 4.5-sized endotracheal tube for deployment. Therefore, bronchial blockers are generally restricted to children who are at least 6–12 months of age. A 3 Fr or 4 Fr Fogerty balloon can similarly act as a useful bronchial blocker in smaller infants but is discouraged because the low-volume, high-pressure properties of the balloon can theoretically damage the airways with prolonged occlusion. In our experience, bronchial blockers can be difficult to position properly within the mainstem bronchus, can often become displaced, and thus are not an ideal option for lung isolation.

Double lumen tubes can be an excellent choice for lung isolation since they provide contralateral ventilation and ipsilateral balloon occlusion. Unfortunately, these devices generally cannot be used in children less than 8–10 years of age (about 25 kg) since the smallest tubes require a 26 Fr airway.

The management of postoperative pain after a thoracotomy incision should be discussed with the anesthesiologist prior to the operation. The options include patient-/nurse-controlled anesthesia, thoracic epidural catheters, pleural catheters, and intermittent IV dosing of pain medications, depending on the type of incision and the age of the patient.

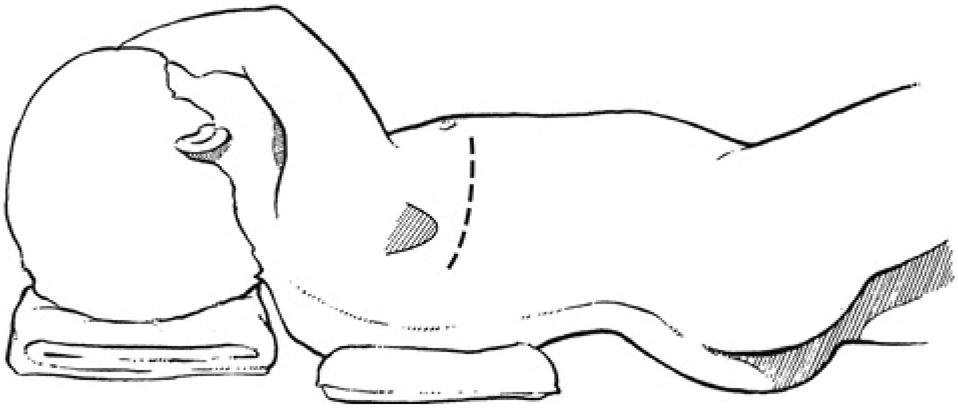
OPEN SURGERY

Posterolateral thoracotomy

Because children have more compliant ribs and are relatively short along their longitudinal axis compared to their adult counterparts, visualization of the pediatric hemithorax through a traditional posterolateral thoracotomy incision is generally excellent.

Position of patient and incision

The child lies in the full lateral decubitus position. In older children, exposure of the intercostal spaces can be enhanced by positioning the patient's iliac spine over the point of flexion on the table. The ipsilateral arm is rotated anterior and cephalad above the nipple. An axillary roll may reduce iatrogenic injury to the brachial plexus, particularly in longer cases (**Figure 8.1**). The child is firmly stabilized in position with towel rolls, gel padding, or a vacuum-type bean bag at the front and back of the chest and abdomen. Care is taken not to create any obstruction or impedance of the incision on range of motion during thoracoscopic approaches. Adhesive strapping is attached across the hips to the operating table and similarly across the uppermost arm and shoulder. The lower leg is well padded and should be flexed at the knee. The upper leg should be straight.

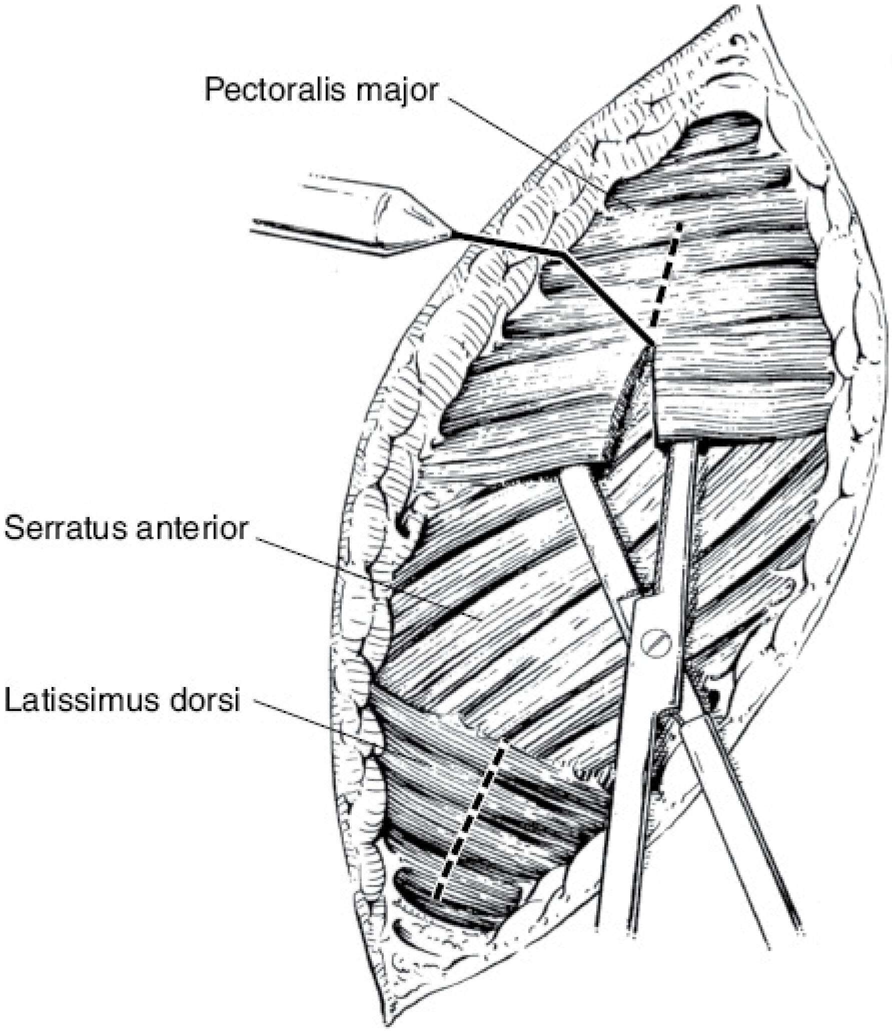


8.1

The skin is widely cleansed with antiseptic solution. Sterile towels are placed to keep the nipple, lower scapula, spine, and costal margin visible as landmarks. The position of the scapula tip as well as nipple should be marked. A large adhesive plastic sheet can be applied to stabilize the towels and reduce heat loss. The lower chest wall is included in the operative field for later chest drain placement, if required.

The transverse skin incision starts around the mid-axillary line just below the level of the nipple. In prepubertal girls, the skin incision should be kept well away from the nipple to avoid scarring of the underlying mammary tissue. The incision extends posteriorly about 1 cm below the inferior tip of the scapula. In teenagers and young adults, an adult-type posterolateral thoracotomy incision that extends posterior to the latissimus dorsi may be required. Rarely, the incision may have to be gently curved further along a line that bisects between the vertebral column and the posterior aspect of the scapula. The subcutaneous tissues are divided.

In the traditional posterolateral thoracotomy, the latissimus dorsi muscles and the serratus anterior muscle are divided. Artery forceps are used to elevate each muscle while it is incised with diathermy. Posteriorly, the latissimus dorsi muscle can be transected as far as the erector spinae muscle without concern for causing iatrogenic scoliosis. Bleeding vessels are accurately coagulated using fine-toothed forceps. The long thoracic nerve runs near the anterior border of the serratus anterior muscle and must be protected. Should it be necessary to divide the nerve, this can be done as far caudally as possible to minimize the effect of a winged scapula. If necessary, the pectoralis major may be incised (**Figure 8.2**).



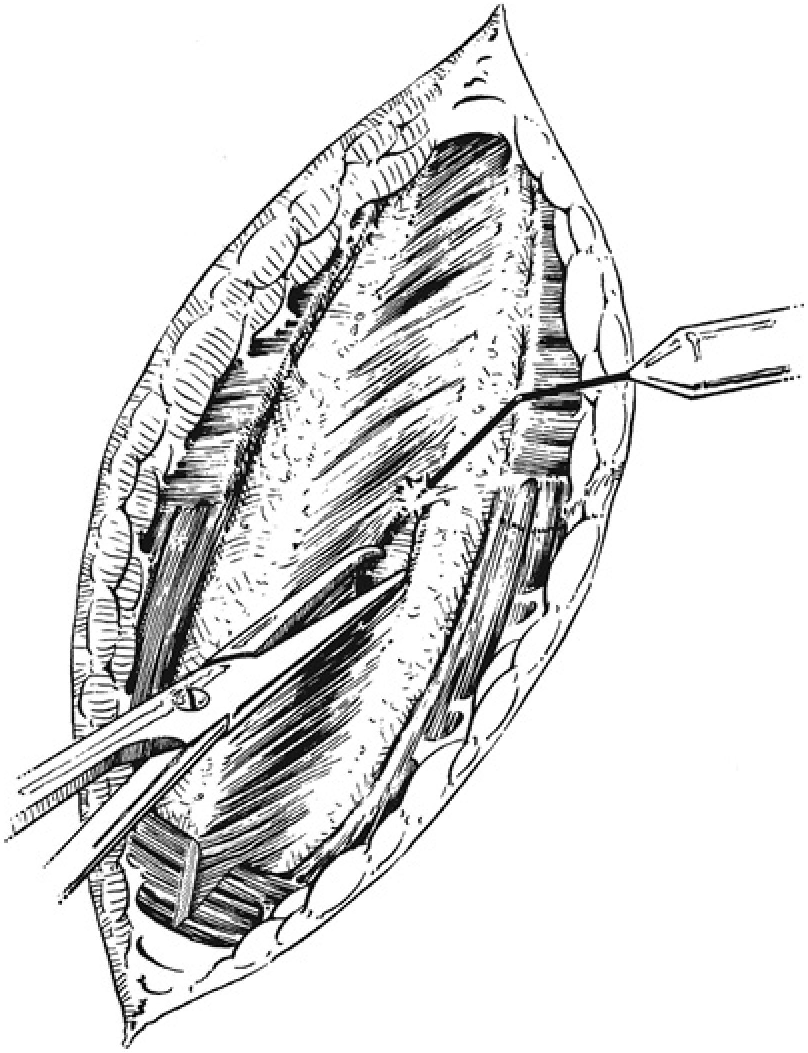
8.2

Once the intercostal muscles are identified, the scapula is elevated with a retractor. The ribs are palpated and counted. In an older child, the highest rib that can be palpated is usually the second rib, on which the posterior scalene muscle inserts laterally. In an infant, the first rib can usually be palpated. The target rib is marked with the diathermy. The fifth intercostal space provides adequate exposure for most pediatric thoracic procedures, including those involving the diaphragm. A key exception is esophageal atresia in which the fourth intercostal space is optimal.

Opening the chest

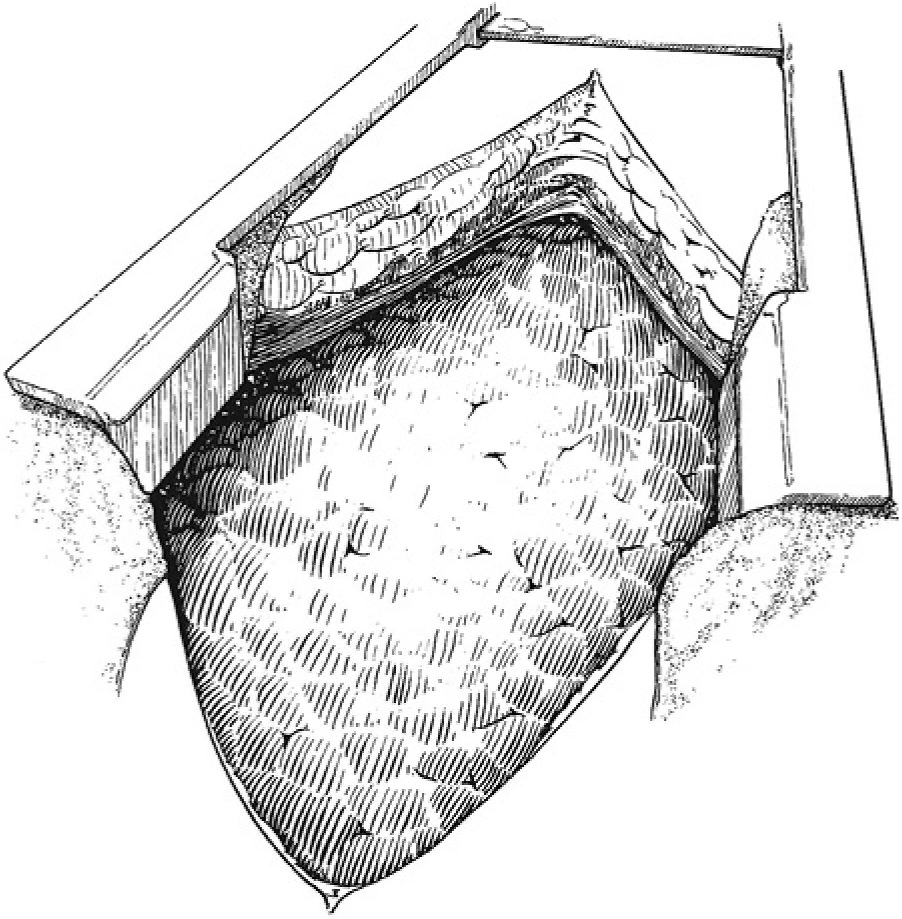
INTERCOSTAL TRANSPLEURAL APPROACH

The fifth intercostal space is entered by dividing the intercostal muscle along the superior aspect of the sixth rib with diathermy (**Figure 8.3**). This avoids injury to the neurovascular bundle that lies just inferior to the rib in a recessed groove. A short incision is made initially, using artery forceps to spread the muscle and with the ipsilateral lung deflated by the anesthesiologist. The remaining intercostal muscles are divided, using a peanut swab or similar retractor to protect the lung.



8.3

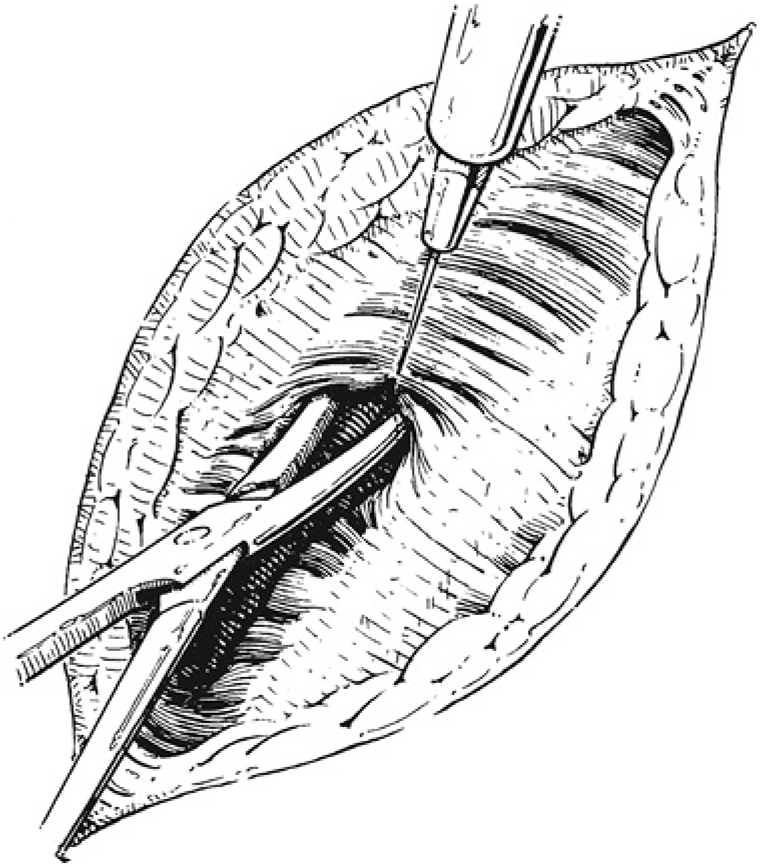
The rib retractor (such as the Finochietto retractor) is inserted and opened slowly while dividing the intercostal muscles anteriorly and posteriorly until there is visualization of the longitudinal fibers of the sacrospinus muscles (**Figure 8.4**). Care must be taken to avoid excessive pressure from the retractor as this may fracture the ribs posteriorly, particularly in infants. Because the ribs are more compliant in children, excision of a subperiosteal segment of the rib, often referred to as shingling, is not necessary. The chest wall muscles and subcutaneous tissues may also need to be incised further to ease the tension on the ribs.



8.4

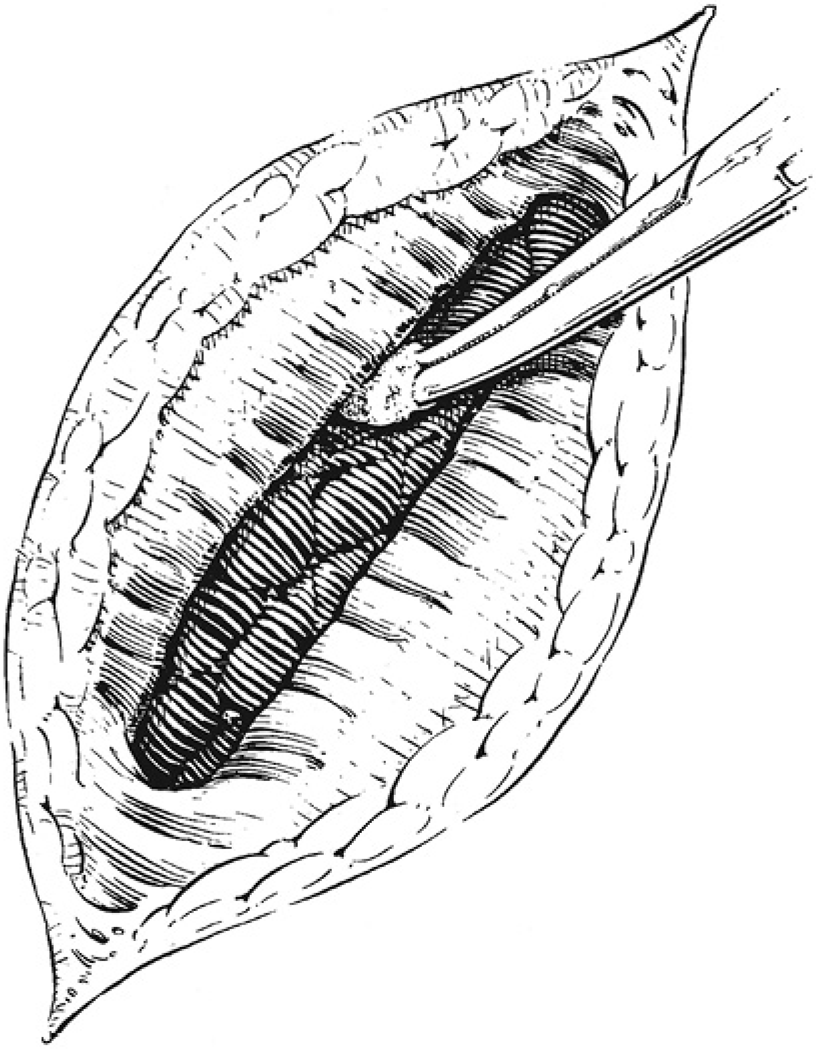
INTERCOSTAL EXTRAPLEURAL APPROACH

Division of the intercostal muscles begins as above using artery forceps to enter the extrapleural space in the posterior half of the incision and diathermy to incise the muscle (**Figure 8.5**). Great care must be taken not to open the parietal pleura.



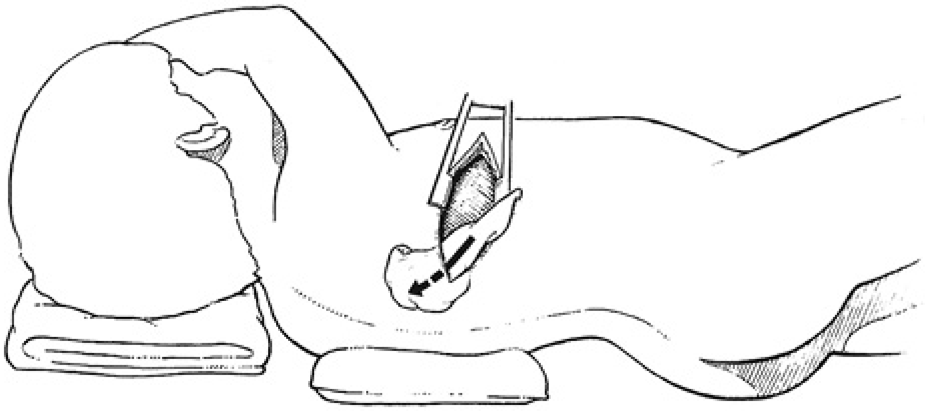
8.5

A moist cotton-tip applicator is used carefully to extend the dissection in the extrapleural plane (**Figure 8.6**). The initial dissection should proceed in a posterior direction. Anteriorly, the parietal pleura is adherent to the ribs and can be easily torn.



8.6

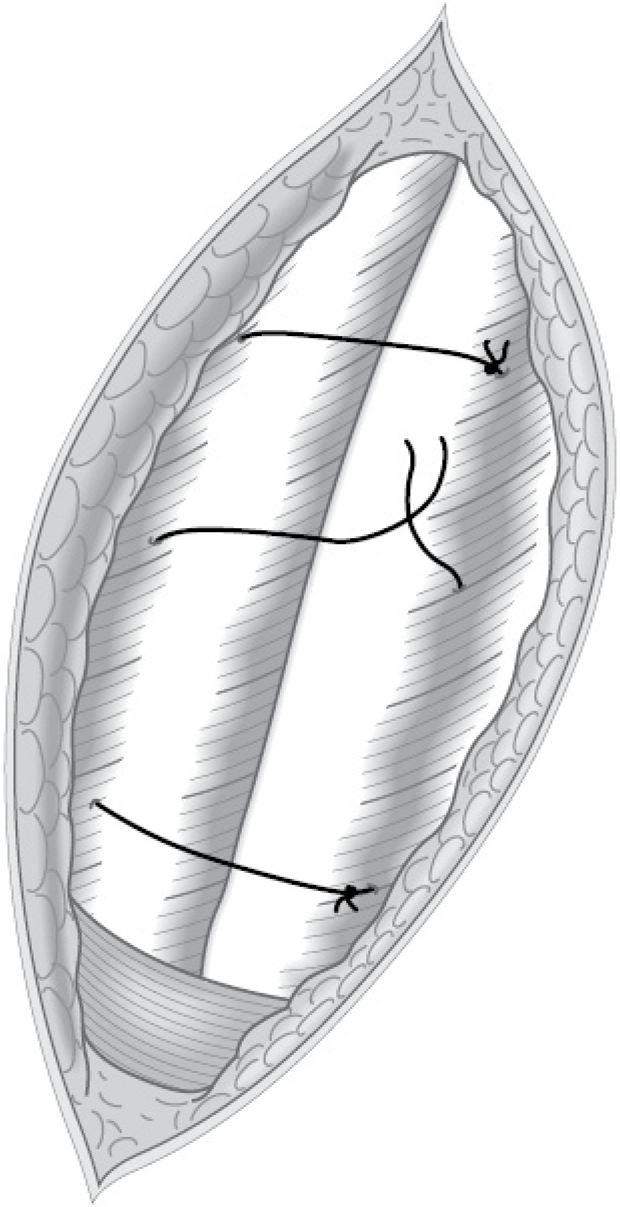
A cotton-tip applicator is used to further develop the extrapleural space posteriorly and superiorly in order to further strip the pleura from the chest wall (**Figure 8.7**). The rib retractor is re-adjusted, and the extrapleural dissection to the posterior mediastinum is completed under direct vision. A moistened gauze sponge can also be used to facilitate this dissection.



8.7

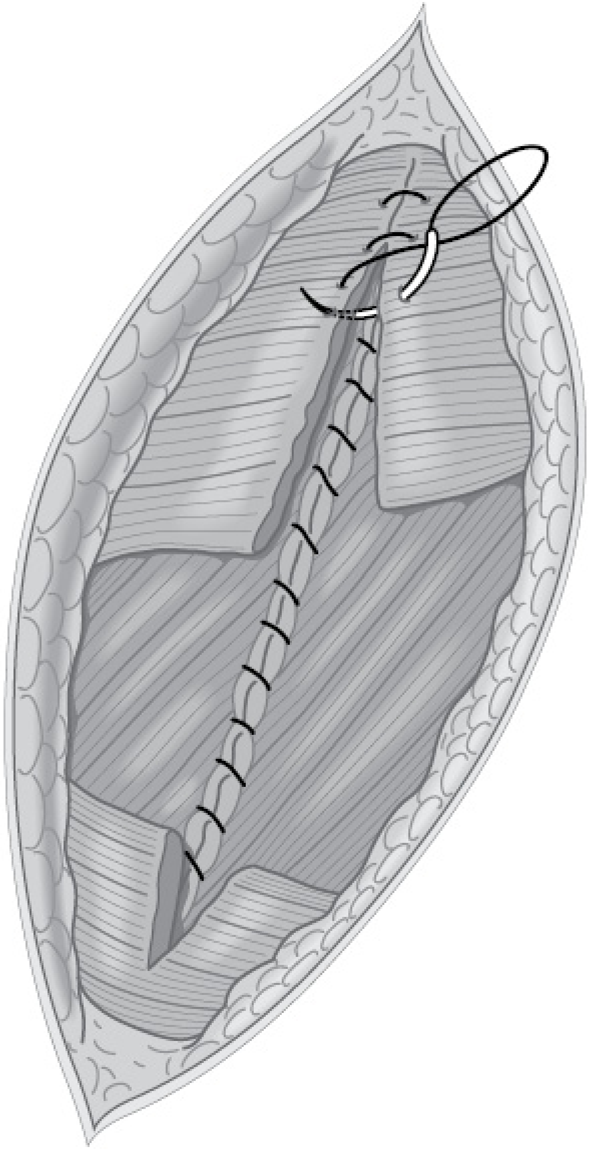
Wound closure

Before closing the chest, posterior rib blocks with bupivicaine above and below the level of the thoracotomy should be performed. Between two and four pericostal sutures are passed around the ribs above and below the incision, taking care to avoid the intercostal vessels (**Figure 8.8**). In infants, an absorbable suture is used and the ribs are loosely approximated such that the normal intercostal space is preserved. Tight apposition of the ribs is to be avoided to prevent rib fusion that may lead to increased pain and other morbidities. Repair of the intercostal muscles is not essential.



8.8

Any divided chest wall muscles are accurately realigned and approximated in anatomic layers using continuous absorbable sutures. The subcutaneous tissues and skin are closed in layers to provide a water-tight seal (**Figure 8.9**). Additional local anesthetic can be infiltrated into the subcutaneous tissues.



8.9

Postoperative care

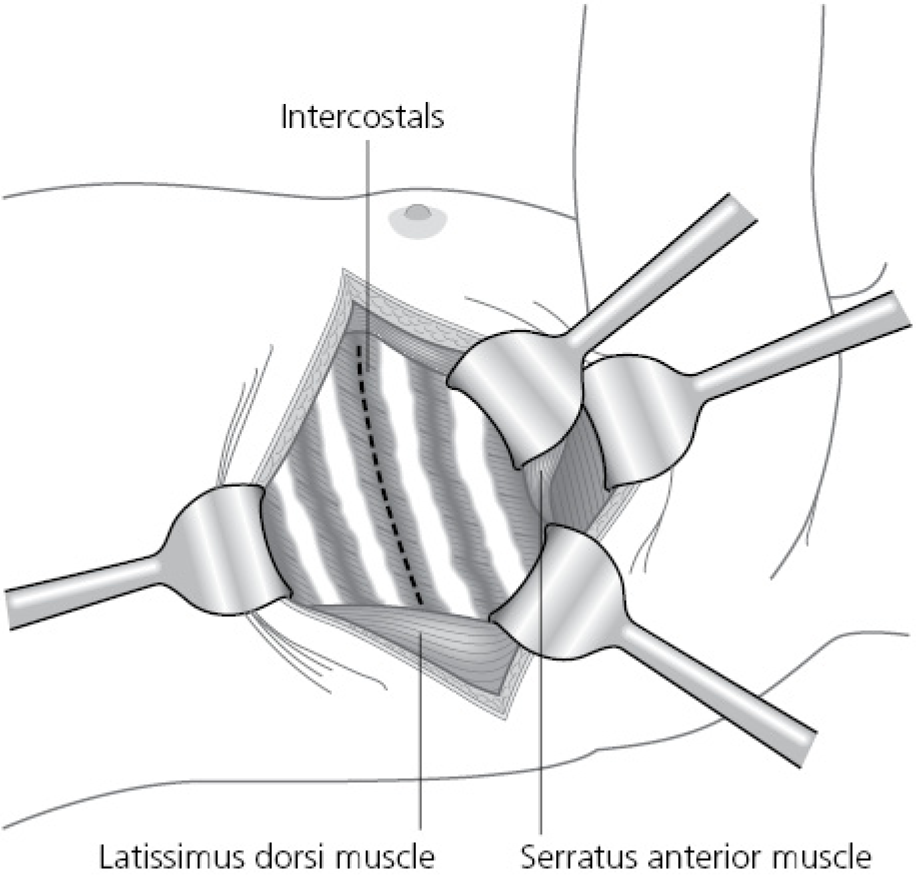
Effective systemic analgesia is essential for the patient's comfort and to facilitate early postoperative physiotherapy and mobilization, which will reduce the risk of atelectasis and pulmonary infections. The use of IV and/or oral non-steroidal inflammatory medications can be effective adjuncts in pain management.

Complications

Early complications include intercostal vessel bleeding and wound infection. Damage to the long thoracic nerve results in winging of the scapula. If the incision is too close to the inferior angle of the scapula, adhesions to the scapula may develop, leading to limited shoulder abduction and elevation. Overzealous reapproximation on the ribs can lead to chest wall deformities and scoliosis. In infants, all follow-up evaluations should include an examination of the spine to screen for scoliosis.

MUSCLE-SPARING THORACOTOMY

The muscle-sparing thoracotomy approach is an excellent alternative to the traditional posterolateral thoracotomy in children. Preservation of the chest wall muscles may reduce postoperative pain and other morbidities. The same skin incision is utilized, and the dissection is carried down through the subcutaneous tissues. Following this, superior and inferior skin flaps are raised, leaving the fascia intact on the muscle. The thin fascial attachments along the anterior border of the latissimus dorsi are incised with diathermy to facilitate retraction of the muscle posteriorly. The fascia along the inferior border of the serratus anterior is incised. The serratus anterior is retracted both anteriorly and superiorly to expose the intercostal muscles (**Figure 8.10**). Occasionally, the lowermost origins of the serratus anterior limit the exposure and can be divided with diathermy. The thoracic cavity is entered through the fifth intercostal space, and the dissection proceeds as described above.



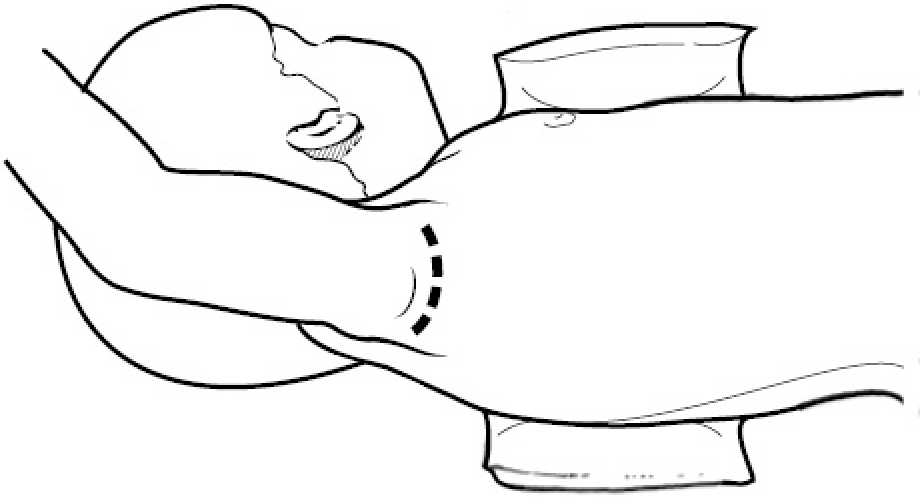
8.10

AXILLARY THORACOTOMY

Axillary thoracotomy is another approach to thoracic access, particularly when visualization of the upper mediastinum is desired. This incision gives excellent exposure of the lung apex, ductus arteriosus, sympathetic chain, and upper esophagus through either the third or fourth intercostal spaces. Axillary thoracotomy can be performed without dividing any major muscle groups and leaves a less conspicuous scar when compared to posterolateral thoracotomy.

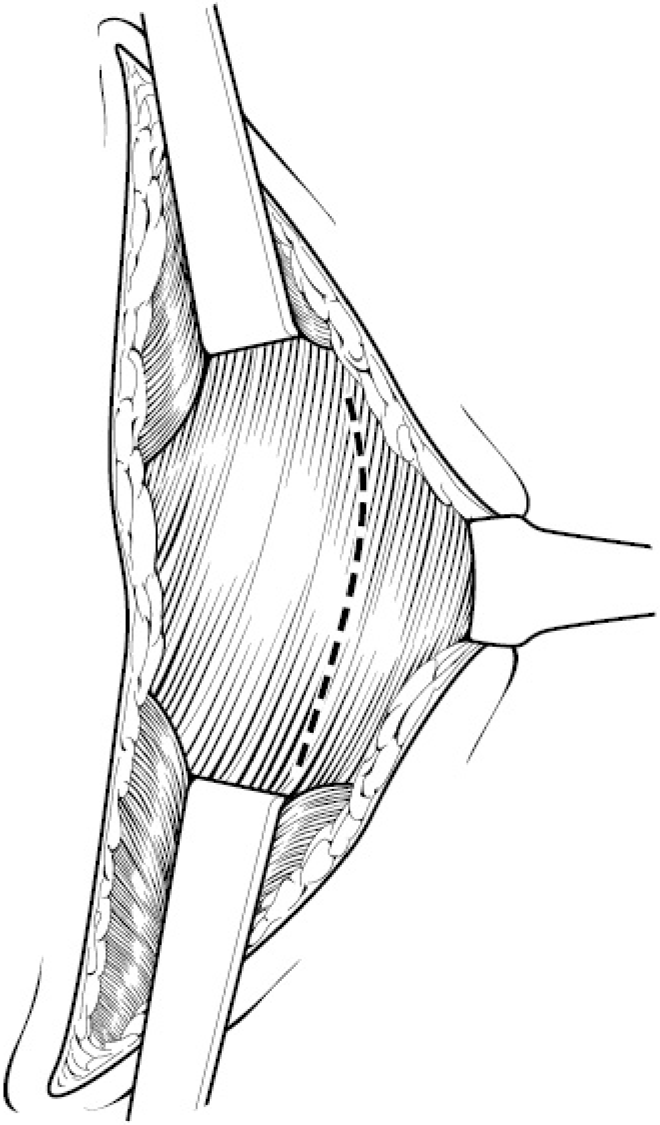
With the infant in the lateral decubitus position and the arm abducted at least 90 degrees above the head, a curvilinear incision is made in a natural axillary skin crease, extending from the lateral border of the pectoralis muscles to the posterior axillary fold formed by the latissimus dorsi muscle

(**Figure 8.11**). A vertical incision in the mid-axillary line down to the ninth intercostal space has also been advocated but tends to leave a less cosmetically appealing scar.



8.11

The axillary fat pad and lymphoid tissues are then swept superiorly. The pectoralis and latissimus dorsi muscles are left intact. The serratus anterior is split transversely along the line of its fibers, taking care to avoid injury to the long thoracic nerve (**Figure 8.12**). Alternatively, the serratus anterior can be mobilized posteriorly and retracted anteriorly to preserve the long thoracic nerve. The thoracic cavity is entered through the third or fourth intercostal space, and the dissection proceeds as described above.



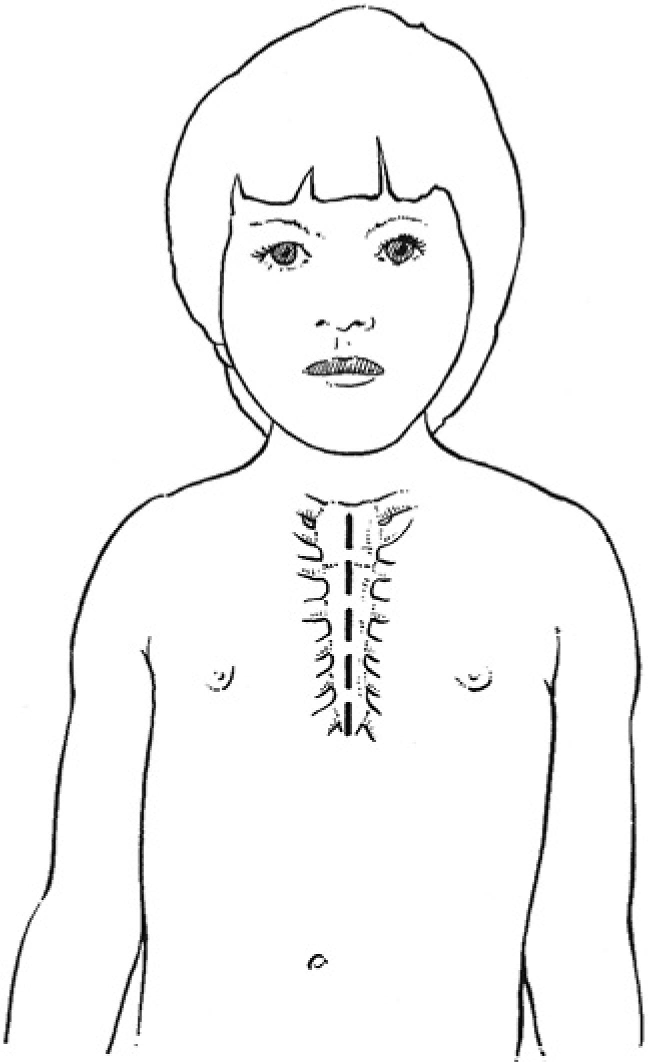
8.12

MEDIAN STERNOTOMY

Although median sternotomy is widely popular in cardiac surgery, this mode of access has a limited role in non-cardiac thoracic operations because visualization of the carina, lower lobes, and diaphragm is generally unsatisfactory. However, a median sternotomy can occasionally be useful when simultaneous access to the bilateral upper/middle lobes or anterior mediastinum (e.g. thymus, upper trachea) is desirable.

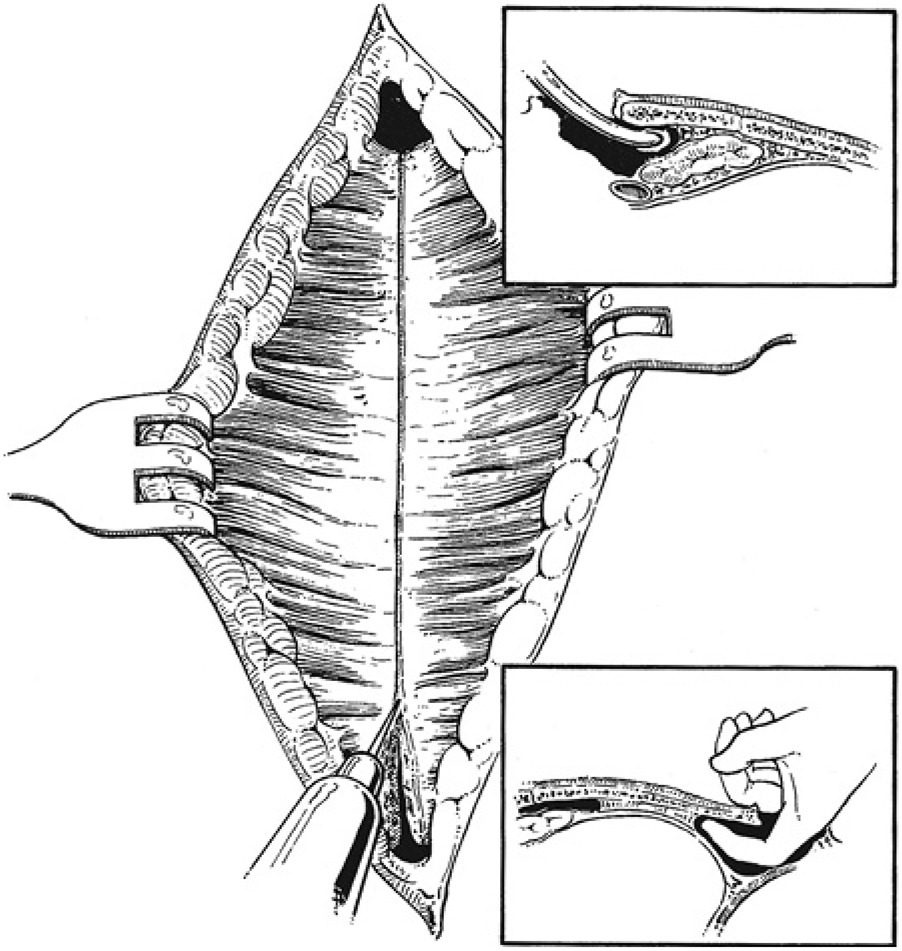
Position of patient and incision

The child lies supine with the arms tucked in and a shoulder roll placed to hyperextend the thoracic spine and bring the sternum forward. Slight elevation of the head of the operating table reduces venous congestion. The skin is incised from just below the suprasternal notch to the xiphoid cartilage (**Figure 8.13**). To minimize blood loss, diathermy is used to incise the subcutaneous tissues and decussating fibers of the pectoralis muscle down to the periosteum.



8.13

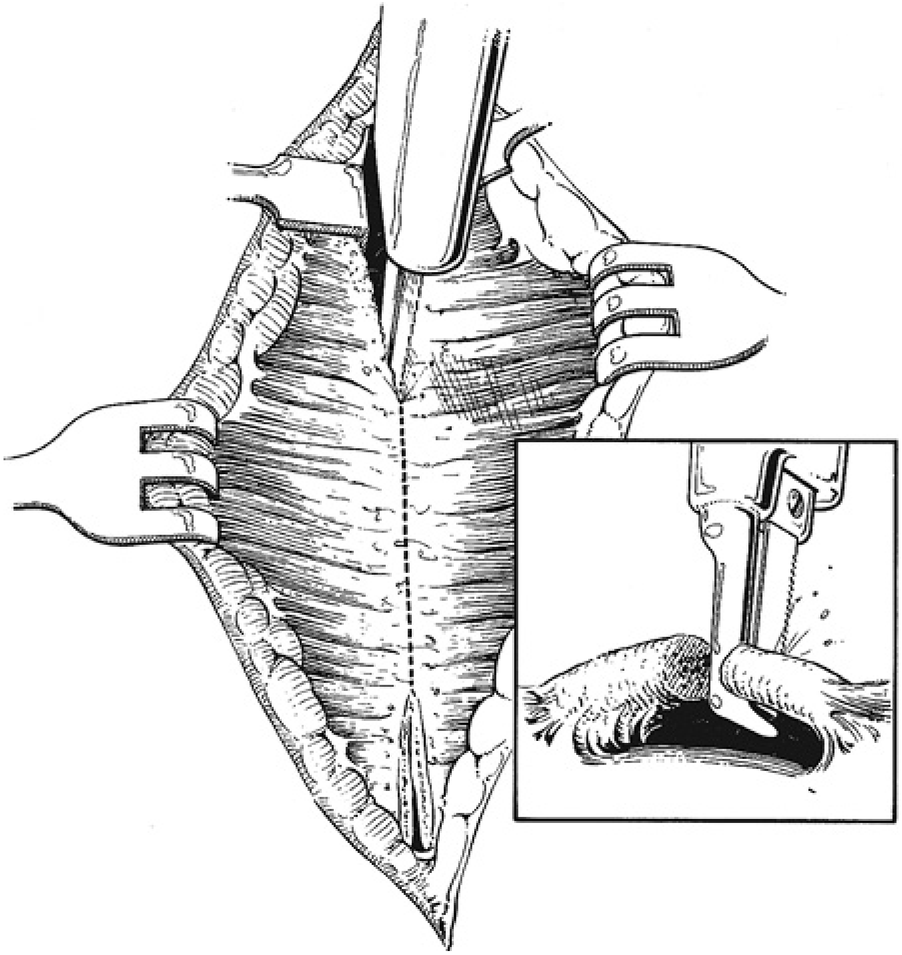
In the suprasternal notch the soft tissues are dissected off the superior and posterior aspects of the manubrium with sharp and blunt dissection. Inferiorly, the xiphoid is mobilized and divided vertically with heavy scissors or diathermy. The retrosternal plane can be entered cranially and caudally using a peanut swab or index finger to free the mediastinal structures from the back of the sternum (**Figure 8.14**).



8.14

Dividing the sternum

The sternum is divided vertically in infants using heavy shears. In older children, a bone saw is required (**Figure 8.15**). Ventilation should be suspended while this is done to avoid opening the pleura. Bleeding, which is usually from the periosteum or bone marrow, is controlled with diathermy and bone wax.



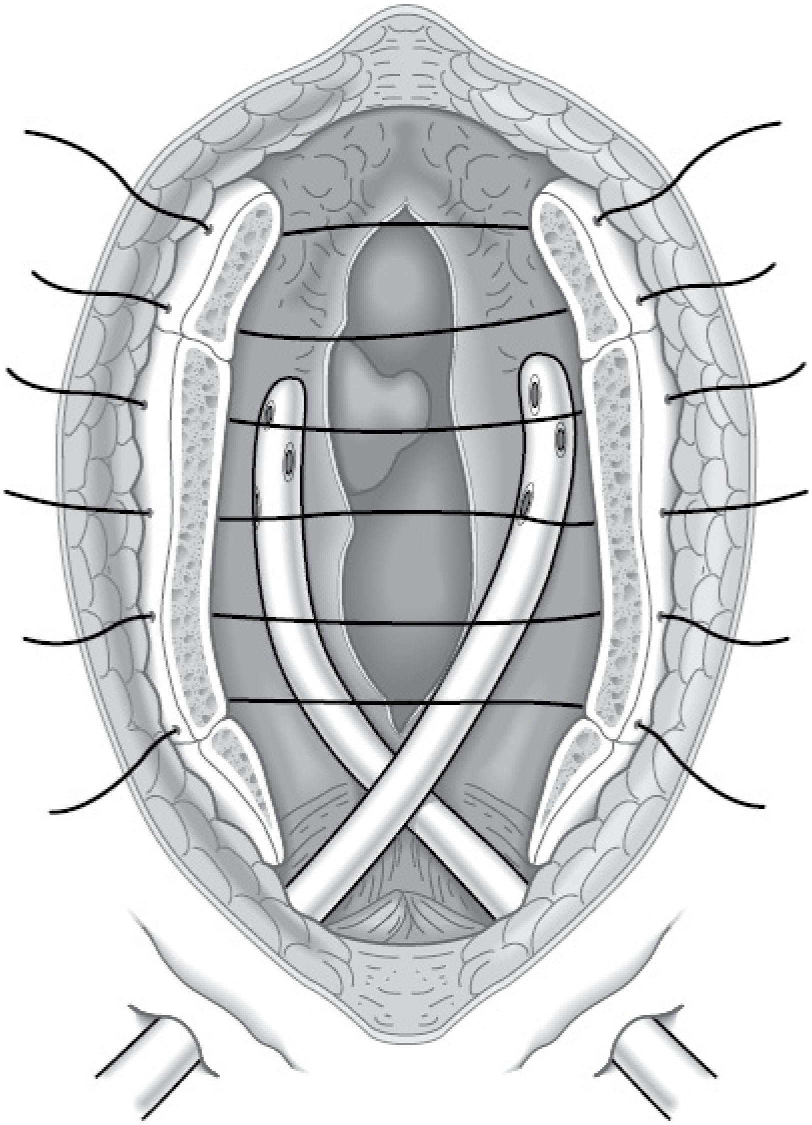
8.15

A self-retraining retractor is inserted and opened slowly while dissecting the soft tissues off the inner aspect of the ribs on either side, taking care not to tear the pleura. In the infant it may be necessary to mobilize and retract or partially excise the thymus gland. This is done by sharp and blunt dissection from below cranially, ligating larger vessels as needed. Laterally, care must be taken not to damage the phrenic nerves.

Wound closure

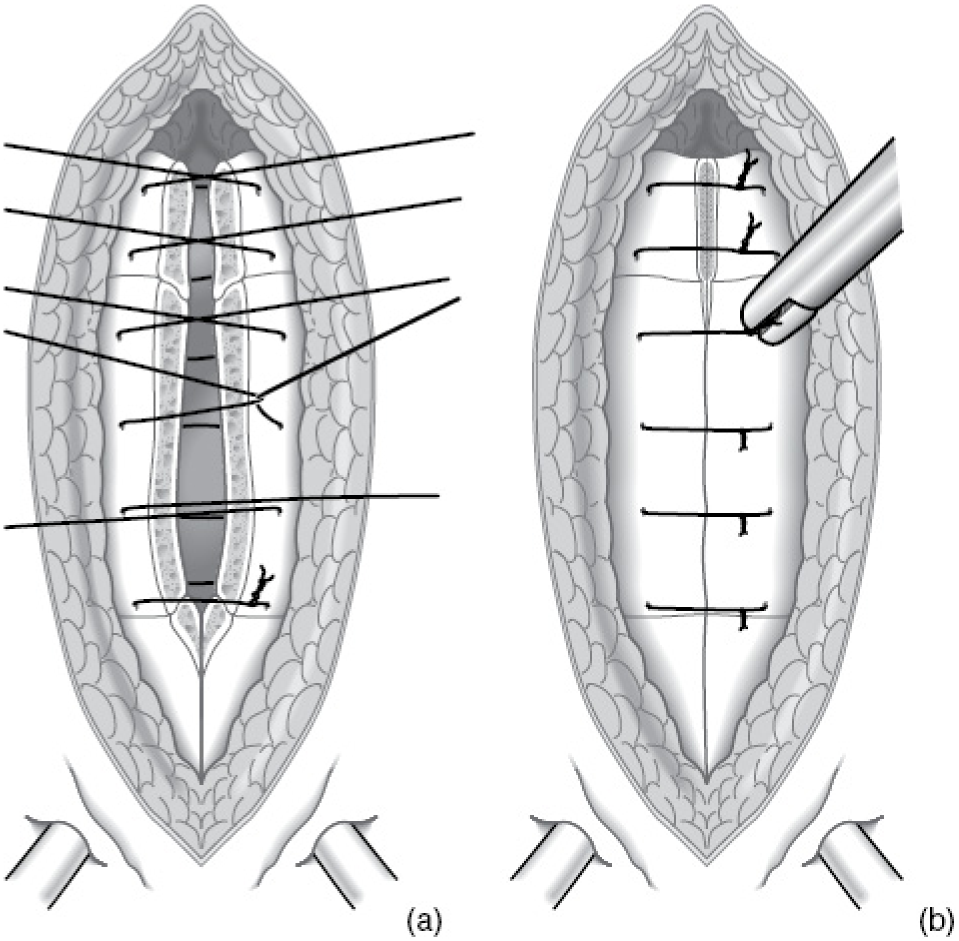
Regardless of whether the pleural cavity has been opened or not, one or two closed suction drainage catheters should be placed in the retrosternal space and brought out through stab incisions below and lateral to the xiphoid process. The drains are sutured to the skin with non-absorbable sutures.

The two halves of the sternum are apposed with no. 2 steel wires (**Figure 8.16**). Heavy sutures (e.g. polydioxanone (PDS), polypropylene) can alternatively be used in infants. In older children, closure with no. 4 or no. 5 wires may be necessary. Wires should be driven through the sternum or taken round the edge of the sternum between sternocostal junctions.



8.16

The ends of the wires are crossed, pulled, and twisted together five to seven times (**Figure 8.17a**). The surplus wire is then cut short and buried into the sternal tissues (**Figure 8.17b**). The subcutaneous tissues are closed with continuous absorbable suture. The margins of the incision are infiltrated with local anesthetic, and the skin is approximated with fine absorbable subcuticular sutures.



8.17a,b

Postoperative care

Systemic analgesia is essential to ensure early mobilization. The drains are removed after 24 hours if no drainage or clinically significant pneumomediastinum is present.

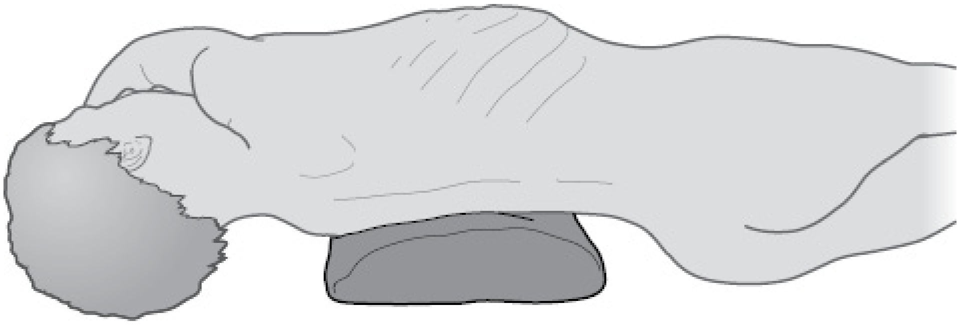
THORACOSCOPIC SURGERY

Minimally invasive techniques have become the preferred operative approach for many thoracic operations, including lung biopsy, pulmonary decortication, and pleurodesis. In the hands of an advanced thoracoscopic surgeon, additional procedures, including diaphragmatic hernia repair, pulmonary lobectomy, and esophageal atresia or tracheoesophageal fistula repair can also be routinely performed in this manner. Thoracoscopy offers a number of advantages compared to traditional thoracotomy, including less pain and better cosmesis. Moreover, the current resolution of high-definition optical systems gives better magnification and therefore superior visualization of the thoracic mediastinum, particularly in the apex. Another potential advantage of minimally invasive techniques is the avoidance of some of the long-term, postoperative morbidity associated with thoracotomy, including limitations in shoulder abduction, chest wall deformities, and scoliosis. Relative contraindications to thoracoscopy include hemodynamic instability requiring pressors, non-conventional ventilation, weight less than 2 kg, and concurrent support on extracorporeal membrane oxygenation (ECMO).

Position of patient and incision

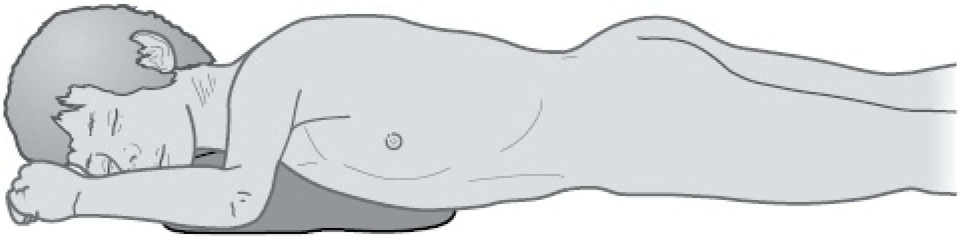
Proper positioning is especially critical for successful thoracoscopy in children. For lobectomy, lung biopsy, and pulmonary decortication, the patient should be in the lateral decubitus position. The same is true for diaphragmatic hernia repair although the infant should be placed crosswise on the operating table to allow the surgeon to stand above the infant's head and preserve appropriate ergonomic alignment between the surgeon, diaphragm defect, and video monitor.

A modified supine position with the ipsilateral thorax elevated about 30 degrees is preferred for biopsy of anterior mediastinal masses, aortopexy, and thymectomy (Figure 8.18). This position allows the lung to retract posteriorly to optimize visualization.



8.18

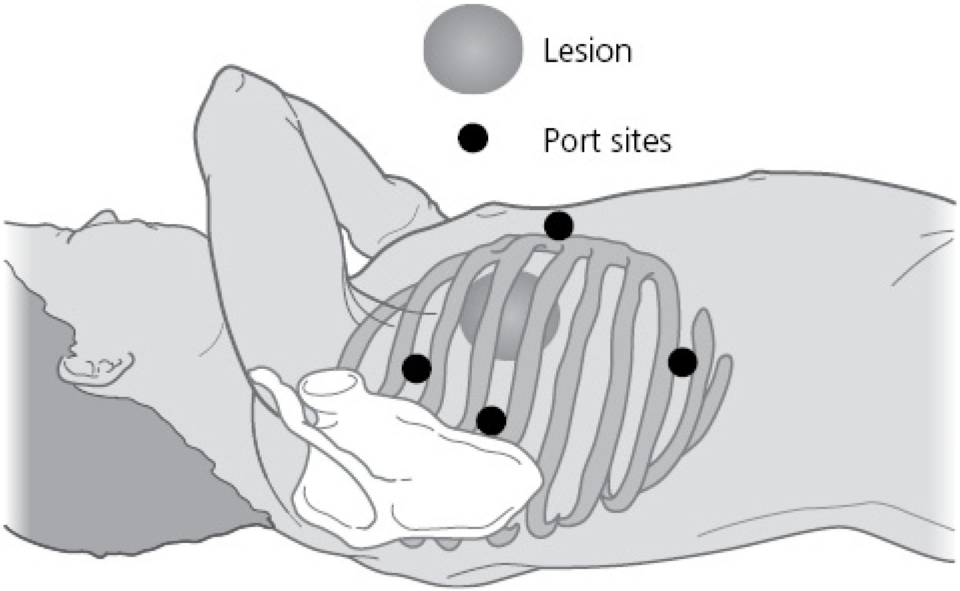
Conversely, a modified prone position is useful for posterior mediastinal masses, such as foregut duplications and neuroblastoma, and for esophageal atresia/tracheoesophageal fistula repair (**Figure 8.19**).



8.19

Once appropriate padding of the child is ensured, the skin is widely cleansed with antiseptic solution and draped since emergent conversion to a thoracotomy is always a consideration. Careful planning of trocar placement is imperative because of the small intrathoracic working space and the potential to damage the thin and delicate thoracoscopic instruments against the rigid chest wall. Common regions for initial entry into the chest cavity are over a rib either posterior to the scapula tip in the "triangle of auscultation" or inferiorly at a proposed chest tube site. After the skin incision is made, the pleural space is accessed with a Veress needle or artery forceps. A 5 mm trocar is gently inserted, and the thorax is insufflated with carbon dioxide to 4 mmHg at a flow rate of 0.1–5 L per second. Higher flow rates can lead to desiccation of the tissues. A 5 mm 30-degree thoracoscope is used to inspect the chest cavity. For all of these positions, the use of gravity by rotation of the bed can be helpful to keep the uninvolved lung and other structures out of the field. If necessary, insufflation pressures of up to 12 mmHg can be tolerated for short periods of time if initial operative visualization is poor.

Many chest procedures require dissection in a focused area within the mediastinum. In these situations, a triangular arrangement of the trocars works best with the camera port placed slightly above and between the two working ports (**Figure 8.20**). This allows the surgeon to look down on the field of view and to minimize instrument dueling. The use of 3 mm or 5 mm ports/instruments is standard in children, depending on the size of the patient. Larger children have wider intercostal spaces that can usually accommodate 12 mm trocars for endoscopic linear stapling. Larger trocars are ideally placed through a lower interspace to maximize working space for the staple head. If the anesthesiologist can effectively establish lung isolation, some surgeons prefer to insert and remove instruments through stab incisions since maintaining positive insufflation pressures within the mediastinum may not be necessary. Upon completion of the procedure, all incisions are infiltrated with local anesthetic and closed in one or two layers using non-absorbable sutures.

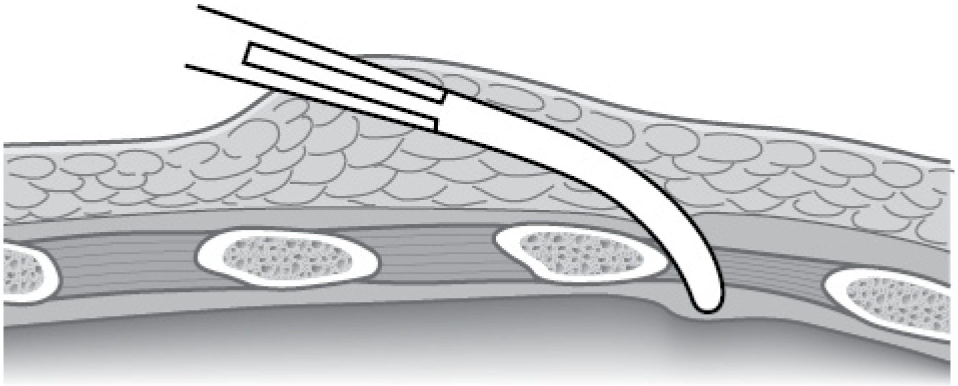


8.20

CHEST TUBES

Any thoracotomy or thoracoscopic procedure in which an air leak or accumulation of pleural fluid is anticipated postoperatively requires placement of an intercostal chest tube at the end of the procedure. Typical tube sizes range from 12 to 28 Fr, depending on the size of the patient and type of pleural drainage anticipated.

Under direct visualization, the chest tube is positioned posteriorly and toward the apex to facilitate drainage and re-expansion of the lung (**Figure 8.21**). The skin incision is ideally placed well away from the thoracotomy incision or through one of the existing trocar sites. An incision adjacent to the costodiaphragmatic recess at least one intercostal space below the site of intercostal entry is preferred to minimize postoperative leakage, particularly in children who have a thin chest wall. The tube should always exit the body anterior to the mid-axillary line extending down to the anterior superior iliac spine. Chest tubes placed posterior to this are more painful and prone to kink. Many chest tubes placed in infants may need to be cut in length to avoiding kinking. Heavy non-absorbable suture is used to secure the tube to the skin followed by application of an occlusive, adherent dressing.



8.21

Depending on the level of concern for a parenchymal air leak, the chest tube should immediately be connected to low-pressure suction (5–15 cm water) or water seal. A chest radiograph to confirm adequate chest tube positioning is mandatory. The use of antibiotic prophylaxis while the chest tube is in place remains controversial. The chest tube should be removed if the lung stays fully expanded on water seal, air leaks have resolved for 24–48 hours, and fluid drainage is minimal, typically defined as less than 2.5 mL per kg per day.

FURTHER READING

Bianchi A, Sowande O, Alizai NK, et al. Aesthetics and lateral thoracotomy in the neonate. *J Pediatr Surg* 1998; 33: 1798–800.

Jaureguizar E, Vazquez J, Murcia J, et al. Morbid musculoskeletal sequelae of thoracotomy for tracheoesophageal fistula. *J Pediatr Surg* 1985; 20: 511–14.

Rodgers BM. The role of thoracoscopy in pediatric surgical practice. *Semin Pediatr Surg* 2003; 12: 62–70.

Rothenberg SS, Pokorny WJ. Experience with a total muscle-sparing approach for thoracotomies in neonates, infants, and children. *J Pediatr Surg* 1992; 27: 1157–9.

Singh SJ, Kapila L. Denis Browne's thoracotomy revised. *Pediatr Surg Int* 2002; 18: 90–2.

Soucy P, Bass J, Evans M. The muscle-sparing thoracotomy in infants and children. *J Pediatr Surg* 1991; 26: 1323–5.

9

Esophageal atresia: Open and thoracoscopic approaches

SHAUN M. KUNISAKI AND STEVEN S. ROTHENBERG

HISTORY

The first description of esophageal atresia (EA) is credited to Durston, who described EA in one of a pair of conjoined twins in 1670. In 1913, Richter proposed the first plan for its management, which comprised dividing the associated tracheoesophagal fistula (TEF) and feeding the infant by gastrostomy until the "technical difficulties of an esophageal anastomosis" had been overcome. Ladd and Leven were independently the first to achieve long-term survival in 1939, but only by a staged approach. Haight in 1941 is credited with the first successful primary repair of EA by fistula ligation and single layer esophageal anastomosis from a left extrapleural approach.

PRINCIPLES AND JUSTIFICATION

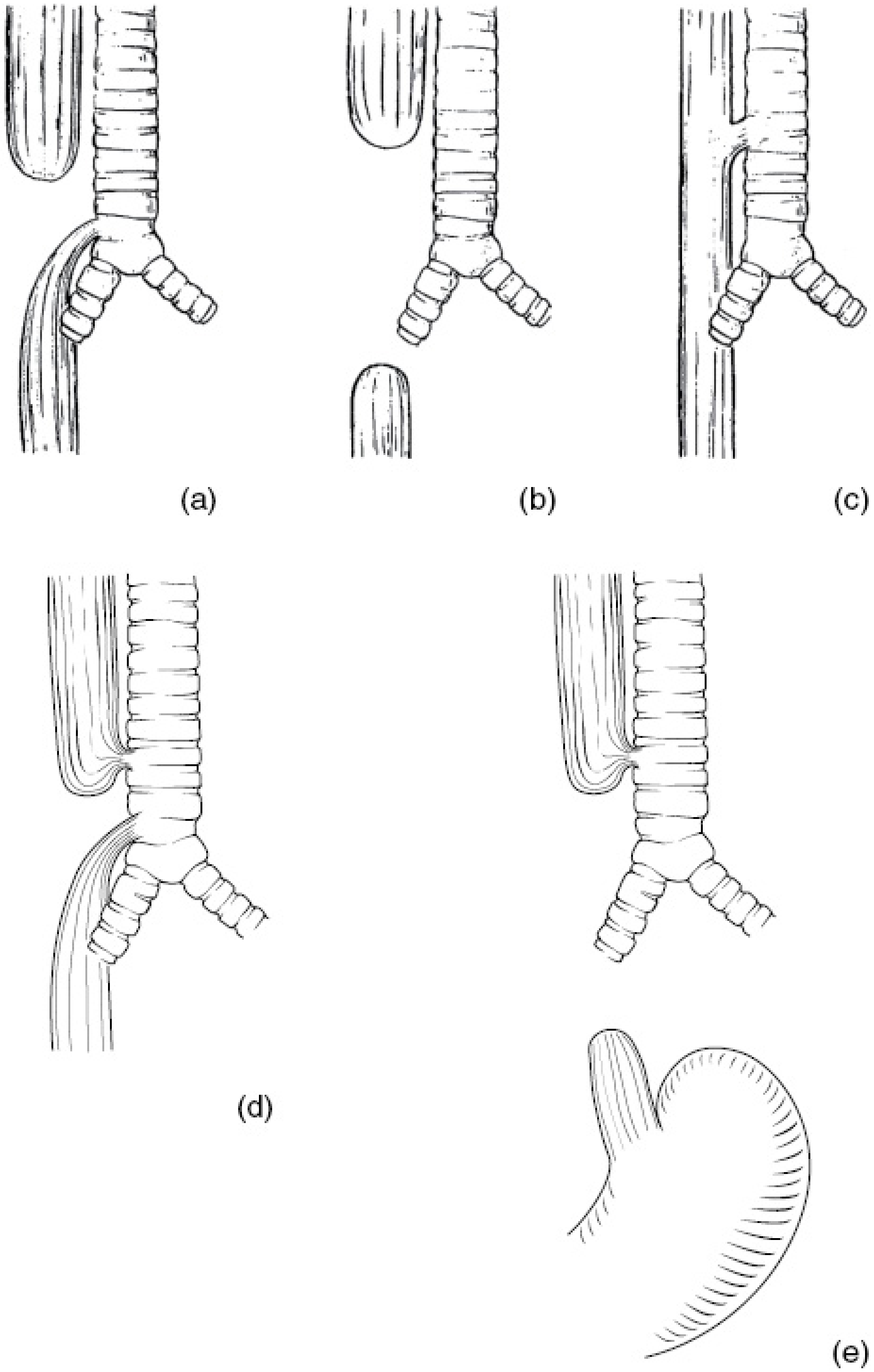
Embryology

The respiratory primordium first appears as a ventral diverticulum of the foregut at 22–23 days' gestation. This is followed by a period of rapid growth when the ventrally placed trachea separates from the dorsally placed esophagus. One theory postulates that the trachea becomes a separate organ as a result of rapid longitudinal growth of the respiratory primordium away from the foregut. An alternative theory is that the trachea initially grows as part of the undivided foregut and then becomes a separate structure as a result of a separation process that starts at the level of the lung buds and proceeds in a cranial direction. This process is associated with a precise temporospatial pattern of expression of several developmental genes, including *sonic hedgehog*, *Sox2*, *Nkx2.1*, and *noggin*. The primary defect resulting in EA is likely the persistence of an undivided foregut either as a result of failure of tracheal growth or of failure of the already specified trachea to separate physically from the esophagus.

Classification of EA

There are various types of EA:

- EA with distal fistula (85%), Gross C (**Figure 9.1a**)
- EA without fistula (6%), Gross A (**Figure 9.1b**)
- H-type TEF (4%), Gross E (**Figure 9.1c**)
- EA with proximal and distal fistula (1%), Gross D (**Figure 9.1d**)
- EA with proximal fistula (4%), Gross B (**Figure 9.1e**)



9.1a–e

Associated anomalies

Over 50% of infants with EA have one or more associated anomalies. The VACTERL association (V, vertebral; A, anorectal; C, cardiac; T, trachea; E, esophageal; R, renal; L, limb) is a well-known combination of defects in which three or more anomalies are present. Accordingly, a full clinical examination should be made at the outset looking for associated anomalies, and special investigations, particularly echocardiography, should be performed prior to repair. The most common defects are cardiac malformations, and these are responsible for the majority of deaths. Next most common are genitourinary tract and vertebral abnormalities.

Prognosis

Unless the child has severe cardiac anomalies, significant chromosomal abnormalities, pulmonary complications, or major prematurity issues, survival is expected with rates now in excess of 90%. The first simplified scheme used to predict mortality was originally proposed by Waterston in the 1960s and updated by Spitz in 1994. Birth weight and cardiac anomalies are major predictors of mortality as shown in Table 9.1. However, more contemporary studies have suggested that birth weight is now less relevant in predicting long-term survival and have proposed alternative models emphasizing the importance of renal anomalies. Morbidity, including dysphagia, gastroesophageal reflux and pulmonary symptoms, continues to be a major problem in many long-term survivors.

Table 9.1 Survival of infants with EA with or without TEF based on birth weight and cardiac anomalies

Class	Birth weight (g)	Major cardiac anomaly	Survival (%)
Low risk	>2000	No	100
Moderate risk	<2000	No	82
Relatively high risk	>2000	Yes	72
High risk	<2000	Yes	27

DIAGNOSIS

Antenatal

Although prenatal ultrasonography is now widespread in most developed countries, the majority of EA cases are not diagnosed antenatally. There is increased suspicion for EA in the presence of a small or absent fetal stomach bubble in combination with polyhydramnios. Nevertheless, the positive predictive value of both ultrasound findings for EA is only 67%. With expert fetal ultrasonography, it is sometimes possible to visualize the blind upper esophageal pouch filling and emptying. The risk of a pure EA is increased when a fetal diagnosis is made.

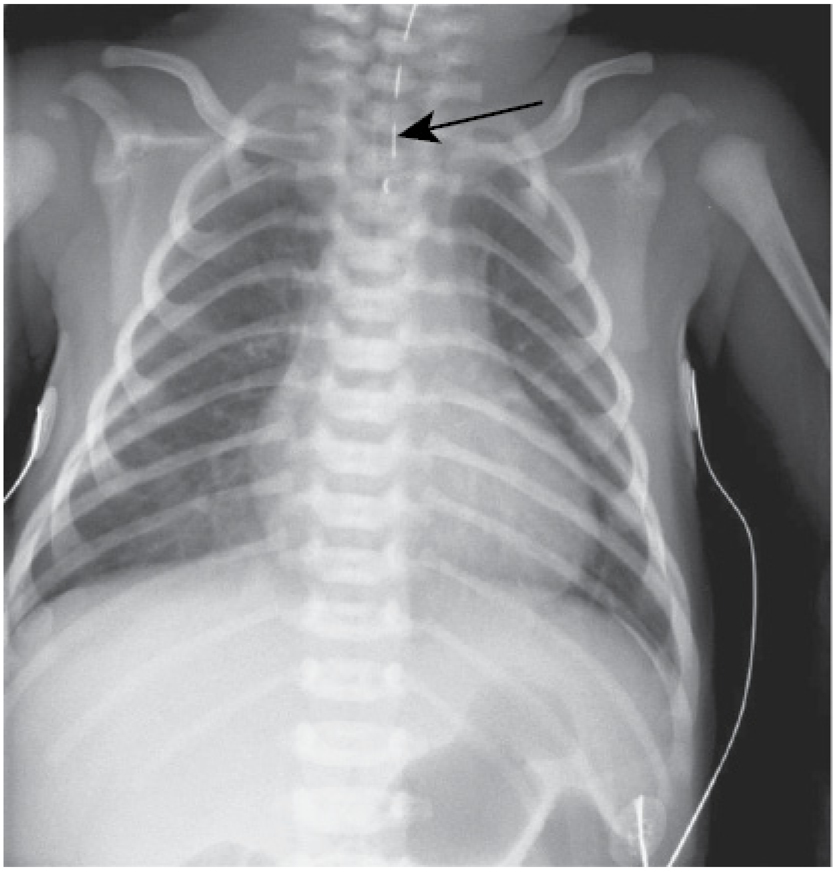
Postnatal

Symptoms that develop in the neonatal period include inability to clear secretions from the mouth, cyanotic episodes with or without attempting to feed the infant, inability to swallow, and respiratory distress. A nasogastric tube (8–10 Fr) should be passed in all symptomatic newborns and in those with increased suspicion based on prenatally polyhydramnios and an absent stomach bubble. Failure to advance the tube beyond 10 cm from the mouth is highly suggestive of EA.

PREOPERATIVE

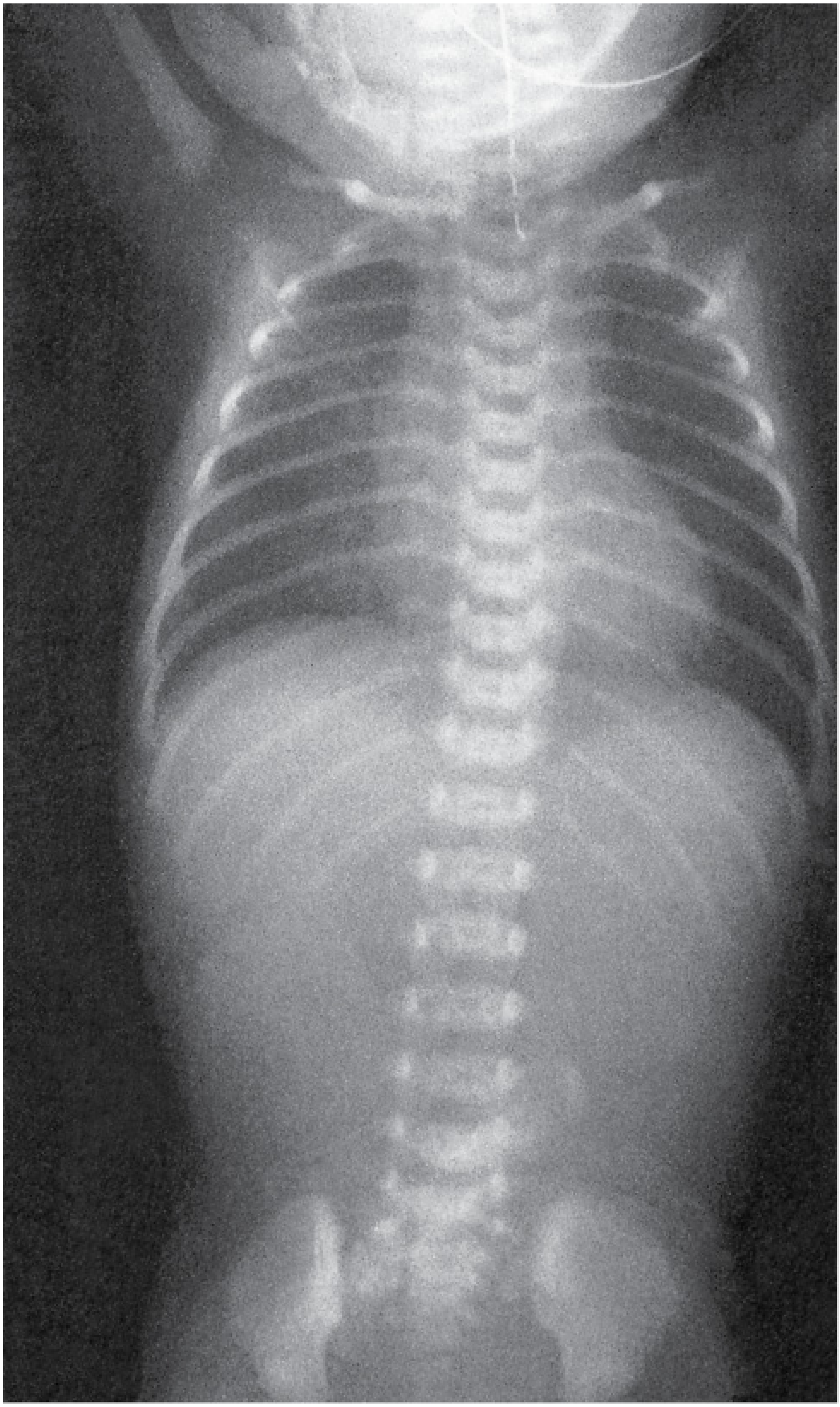
Investigations

In EA, plain radiography of chest and abdomen demonstrates a radio-opaque nasogastric tube coiled in the upper mediastinum. Air in the stomach and intestine suggests the presence of a distal TEF, the most common type of malformation (**Figure 9.2**).



9.2

The complete absence of intestinal air with a coiled nasogastric tube in the upper mediastinum suggests the diagnosis of pure EA (**Figure 9.3**). Less common would be EA with proximal fistula. Contrast studies of the upper pouch can be performed prior to operative repair but rarely provide additional information. If contrast medium is used, the examination should be performed with extreme care by an experienced radiologist to minimize aspiration.



9.3

Initial management

Emergency surgery for EA is seldom necessary. Scheduling the operation 12–24 hours after admission allows for full assessment for associated anomalies and resolution of pulmonary atelectasis if aspiration has occurred prior to referral. While awaiting surgery, the upper pouch is continuously aspirated using a Replogle tube attached to continuous low-pressure suction. Intravenous fluids will maintain fluid and electrolyte balance and prevent hypoglycemia. Broad-spectrum antibiotics are routinely administered preoperatively. An echocardiogram is mandatory to characterize any cardiac defects and to determine the position of the aortic arch.

Neonates with respiratory distress requiring assisted ventilation, particularly if associated with gastric distention, are best approached by urgent transpleural ligation of the distal fistula. This will immediately improve the respiratory status, and gas exchange improves as the escape of gas through the fistula is halted. In some infants, the improvement is so dramatic as to allow primary repair of the atresia to proceed at the same setting. In others, the procedure should be terminated pending improvement in the infant's clinical condition. The esophageal repair can be safely postponed for 7–14 days; further delay increases the risk of the fistula reopening. The rare infant who is in respiratory extremis should have an open gastrostomy tube (10 Fr or 12 Fr Malecot) placed at the bedside to help decompress the gastrointestinal tract. Placement of a vessel loop around the gastroesophageal junction can also be helpful for stabilizing these patients.

Choice of operation

In most neonates with EA and a distal TEF, the aortic arch is left-sided. Therefore, division of the fistula and primary anastomosis of the esophagus should be performed through a right-sided approach. The presence of a right-sided aortic arch, as determined by echocardiogram, occurs in 5% of cases and would indicate a left-sided surgical approach to provide easier access to the mediastinum.

The absence of a lower pouch fistula is usually associated with a long gap between the upper and lower esophagus. This situation should initially be managed by a feeding gastrostomy. In most cases, a delayed primary anastomosis after a short period to allow for spontaneous growth of the pouches is possible. The decision regarding timing of delayed primary anastomosis is based on serial radiological assessment of the intervening gap. One technique for measuring the gap involves placement of a radio-opaque tube into the proximal esophagus and a flexible neonatal endoscope through the gastrostomy stoma and advanced into the distal esophagus. When the gap between the upper and lower esophagus measures less than the height of two vertebral bodies under fluoroscopy, a primary anastomosis (open versus thoracoscopic) should be attempted. Those with longer gap measurements in most cases can still be brought together in a single-stage operation and we have found greater success with a thoracoscopic approach. Some may be candidates for the Foker technique. However, a persistent gap of greater than six vertebrae, especially if there is only a diminutive lower pouch, may indicate the need for an esophageal replacement, but this should be contemplated only after it is clear that a primary esophageal anastomosis is not possible. It is usually not profitable to wait longer than 8–12 weeks before deciding on whether to proceed with delayed primary repair. For older infants with a plan for eventual esophageal replacement, a cervical esophagostomy may be helpful to minimize the risk of aspiration and to allow for sham feeds to prevent oral aversion.

Anesthesia

Although left mainstem bronchus intubation is not required, the endotracheal tube requires careful positioning to permit adequate ventilation with minimal gas flow through the fistula. An IV infusion is sited in an extremity other than the right upper limb.

Preoperative bronchoscopy

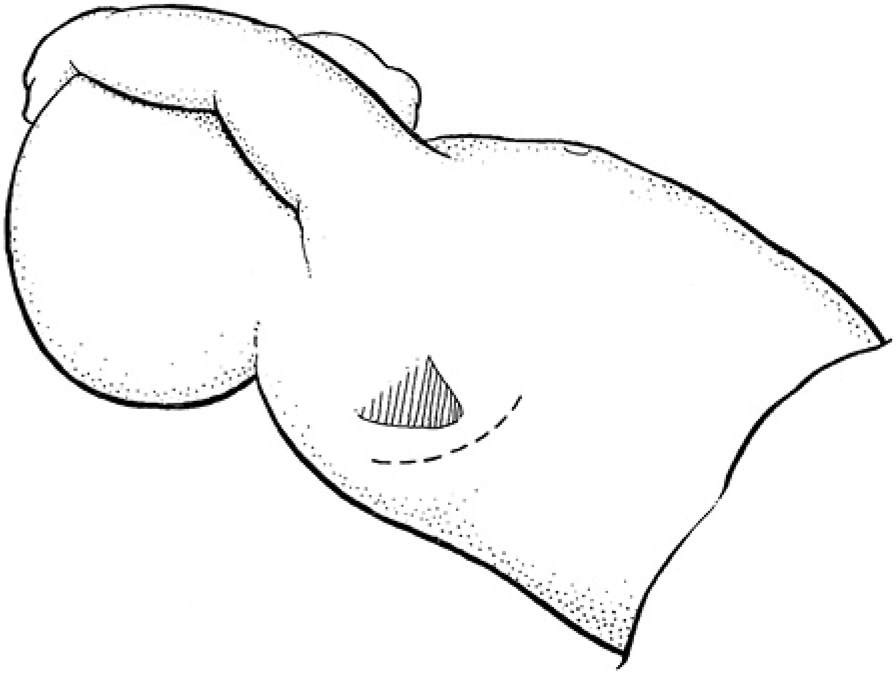
At the time of anesthesia induction, many surgeons advocate rigid bronchoscopy to better define the site of entry of the distal TEF, to look for unrecognized anomalies such as an upper pouch fistula, and to assess the degree of tracheomalacia. A distal TEF entering the trachea at the level of the carina may indicate a longer gap. In cases of suspected pure atresia bronchoscopy should always be performed as the incidence of an upper pouch fistula may be significantly higher than the 4% previously reported.

OPERATIONS

Open (thoracotomy) repair

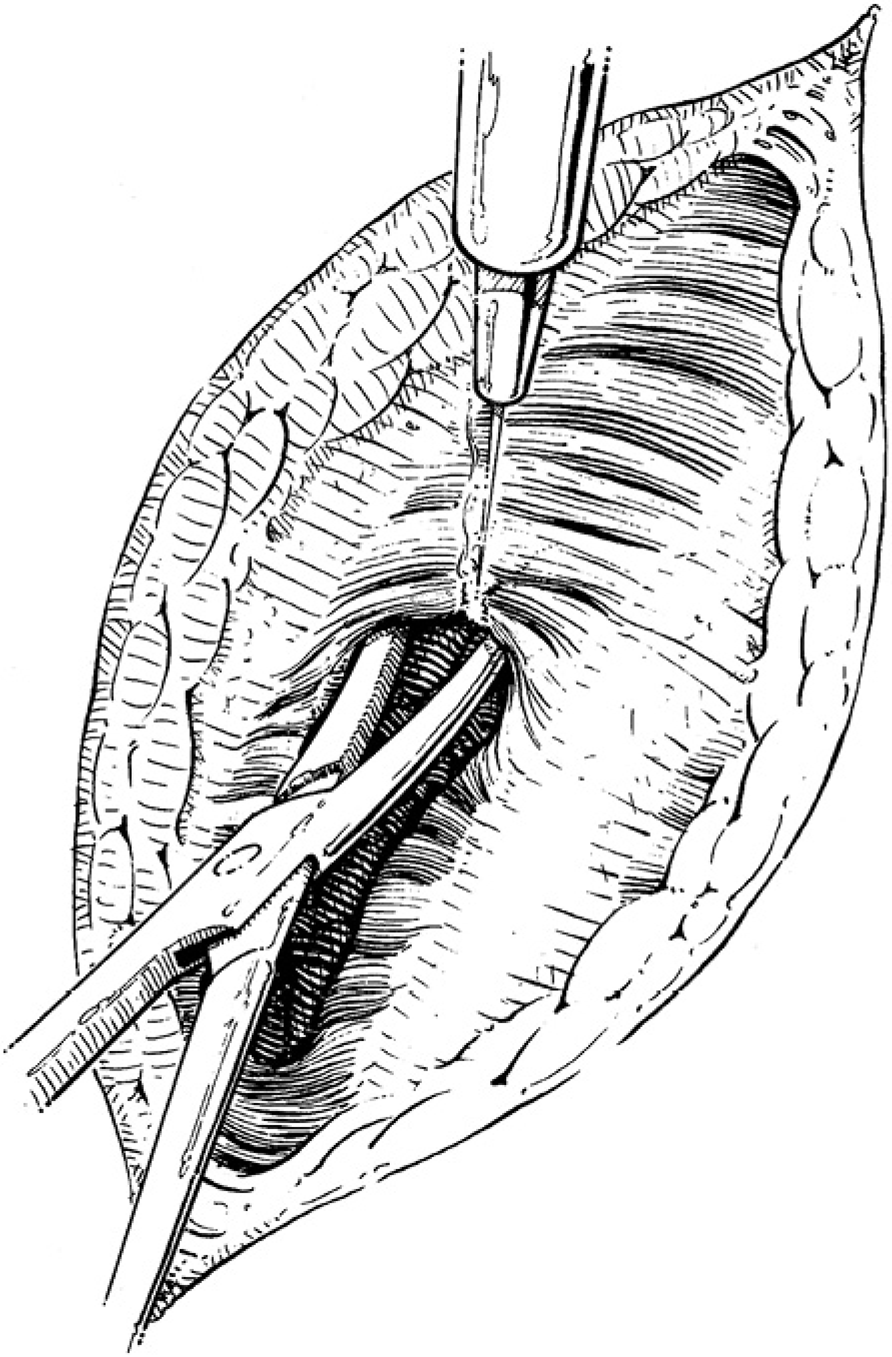
INCISION

For the right-sided approach, the infant is positioned on the left side and stabilized with gel rolls. The right arm is extended above the head and fixed and an axillary role should be placed. A lateral thoracotomy incision is made 1 cm below the inferior angle of the scapula (**Figure 9.4**). Division of the subcutaneous tissues and muscles is carried out with diathermy to minimize blood loss. The lastissimus dorsi muscle is preferably mobilized posteriorly (muscle-sparing approach) or divided along the length of the incision. The serratus anterior muscle is retracted anteriorly, but if additional exposure is required, the lowermost fibers can be divided without compromising its blood supply. An alternative access to the intercostal spaces is through an axillary thoracotomy extending from the lateral border of the pectoralis major muscle to the posterior axillary fold formed by the latissimus dorsi.



9.4

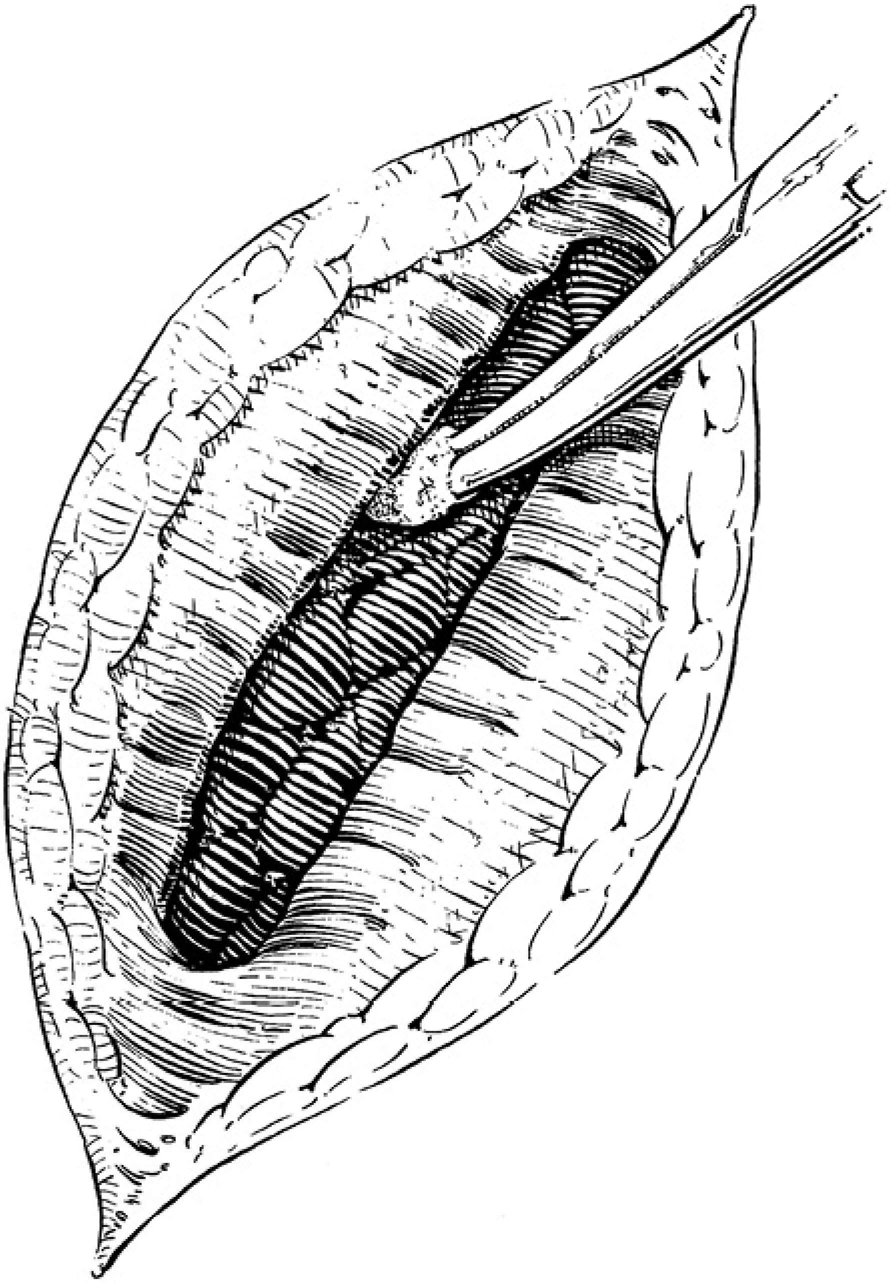
Upon identification of the intercostal muscles, the scapula is elevated and the rib spaces are counted by inspection and palpation. The thorax is best entered through the fourth intercostal space. The external intercostal muscles adjacent to the fifth rib are carefully divided using diathermy (**Figure 9.5**). The internal intercostal muscles are gently elevated and divided from the underlying parietal pleura. The paravertebral fascia and muscles should not be incised.



9.5

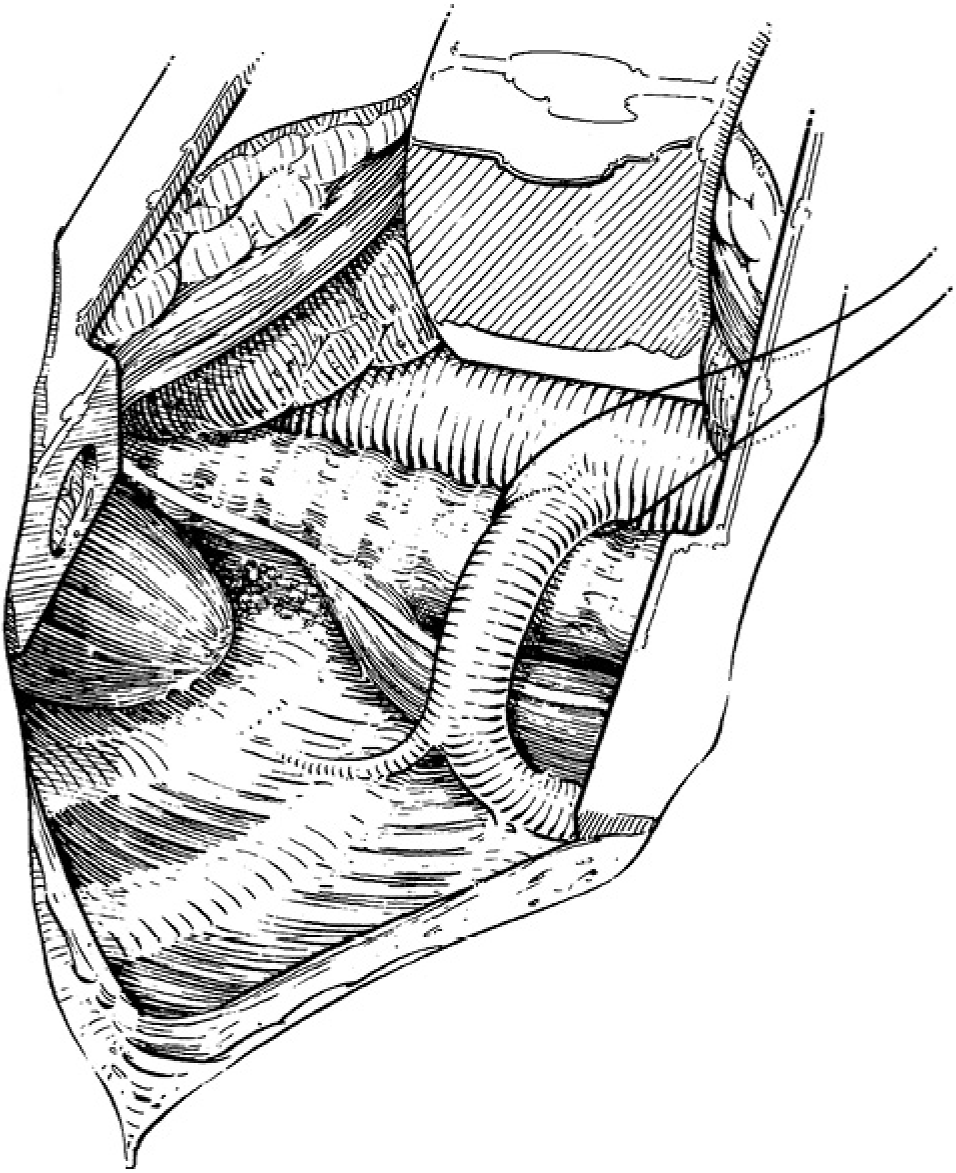
Upon exposure of the pleura, separation of the pleura from the chest wall is best carried out by the gentle dissection with a peanut or cotton-tip applicator (**Figure 9.6**). Once the retropleural space has been developed, a small rib spreader can then be inserted and the ribs gently separated.

Further posterior dissection of the pleura is achieved by using moist gauze or cotton-tip applicators.



9.6

The azygous vein and posterior mediastinum should be exposed, enabling the lower pouch, upper pouch, and fistula to be seen (**Figure 9.7**). Anterior dissection of the pleura should be sufficient only to allow the ribs to be adequately spread. Very occasionally, the size or position of the fistula may make it impossible for the anesthesiologist to ventilate the lungs adequately. In that situation, the more rapid transpleural approach to the fistula may be necessary. In order to expose the posterior mediastinum effectively, lung retraction is essential, but care must be taken to ensure that the retractor does not compress the mediastinal structures.

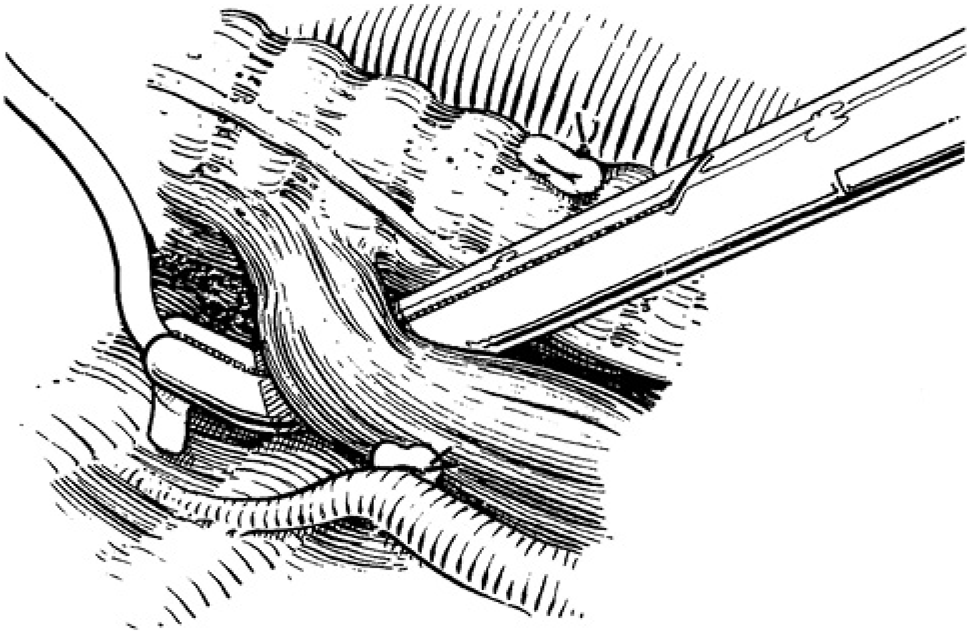


9.7

MOBILIZATION OF THE LOWER ESOPHAGUS AND DIVISION OF THE FISTULA

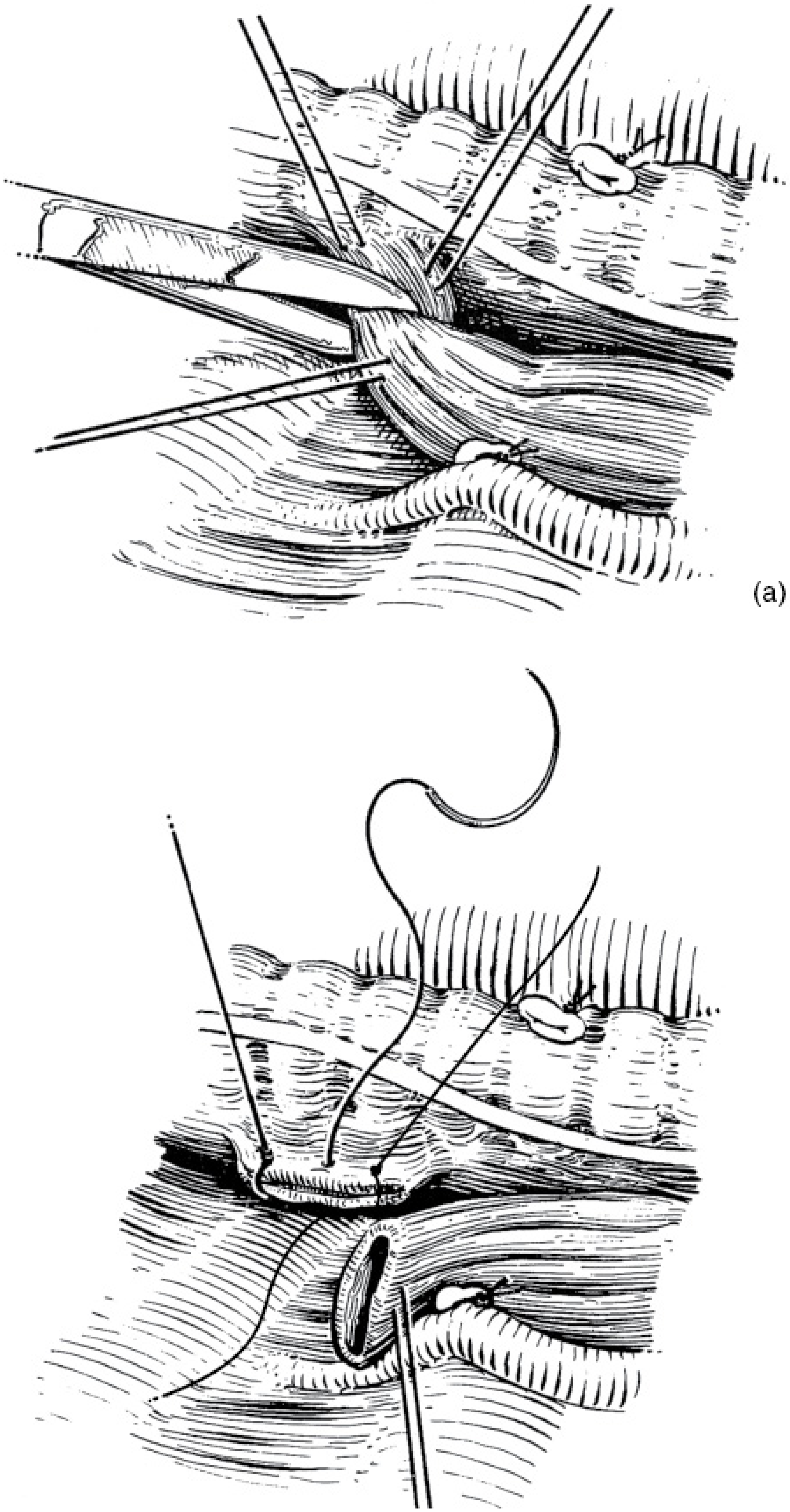
Unless the infant has an interrupted inferior vena cava, the azygous vein can be routinely ligated with 3/0 or 4/0 sutures and divided. Once this is accomplished, the lower esophagus is more readily identified, distending with each inspiration in the lower posterior mediastinum. The close proximity of the vagus nerve to the lower esophagus can also aid in its identification. If the dissection is carried out too posteriorly, the aorta could come into view mimicking the esophagus distal to the TEF.

The lower esophagus is dissected circumferentially to just distal to the fistula and a loop is placed around it (**Figure 9.8**). This maneuver is usually accompanied by an audible sign from the anesthesia team as ventilation becomes easier with fistula occlusion. Traction on this loop controls the fistula and enables the junction of the lower esophagus and trachea to be accurately defined and dissected as needed. In addition, occlusion of the fistula by traction on the loop while inflating the lungs will eliminate the possibility of mistaking the right mainstem bronchus for the distal esophagus.



9.8

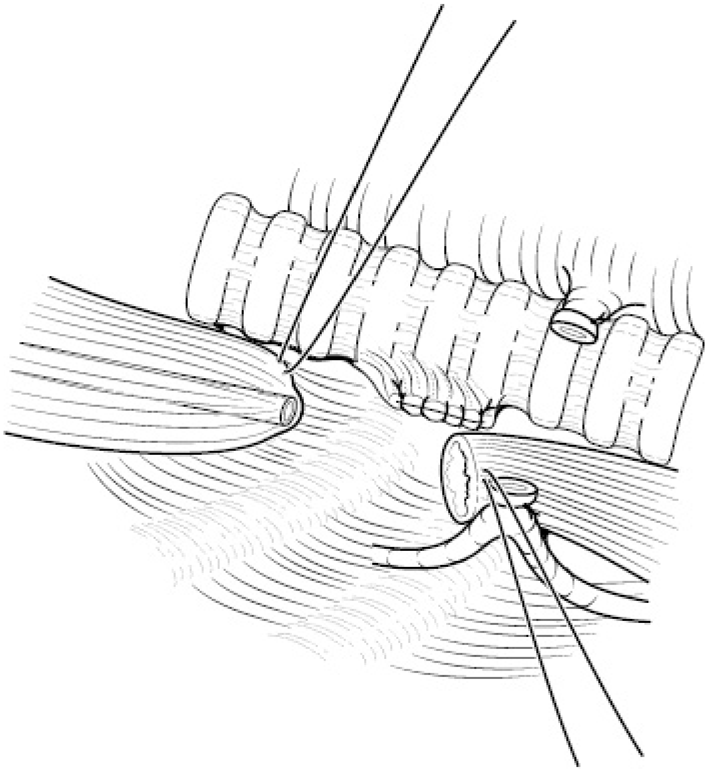
After carefully defining the junction between the trachea and esophagus, two 5/0 or 6/0 absorbable sutures are placed in the trachea at cranial and caudal ends of the fistula, and the fistula is divided a few millimeters distal to its entry into the trachea (**Figure 9.9a**). The trachea is then closed with multiple, interrupted sutures (**Figure 9.9b**). The airtightness of the closure should be tested by instiling saline into the mediastinum and watching for bubbles while ventilating to 30–40 cm H_2O pressure. Prior to dissection of the lower esophagus, a 5/0 stay suture into the lower esophagus may allow for traction to be exerted on the lower esophagus without handling with forceps. Dissection of the lower esophagus needs care to preserve the vagal attachments and to prevent damage to the adjacent thoracic duct and left pleura. Complete mobilization of the lower pouch has traditionally been discouraged (because of concerns about devascularization), but in most cases, lower pouch mobilization may be achieved down to the level of the diaphragm to facilitate less tension on the anastomotic repair.


(a)

9.9a,b

IDENTIFICATION AND DISSECTION OF THE UPPER POUCH

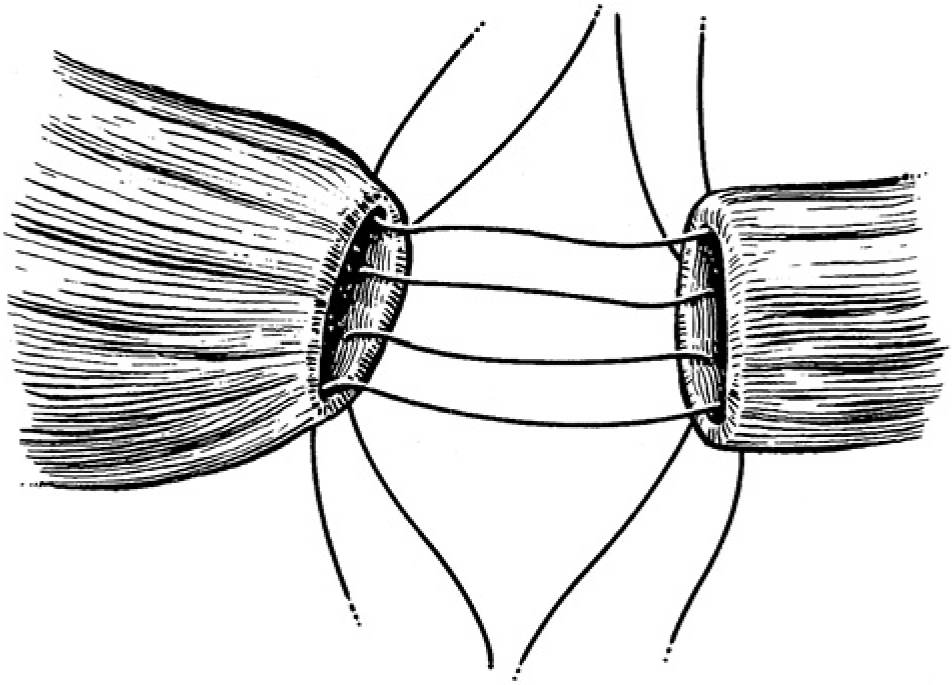
If the upper pouch is not immediately visible, placement of a soft catheter, bougie, or Bakes dilator into the upper pouch by the anesthesiologist will help with its identification. A traction suture can be placed into the tip of the upper pouch to facilitate its exposure and minimize the need for forceps traction (**Figure 9.10**). The proximal esophagus has a rich blood supply coming from the thyrocervical trunk and therefore can be dissected well into the thoracic inlet. Branches of the vagus nerve supplying the upper esophagus as well as the recurrent laryngeal nerves should not be disturbed. Dissection in the common plane between the esophagus and trachea is best carried out sharply and with extreme care to avoid inadvertently opening into either structure. Dissection of the upper pouch is sufficient when an anastomosis can be made with the distal esophagus. The size of the opening in the upper esophagus should correspond to the diameter of the lower esophagus.



9.10

ANASTOMOSIS

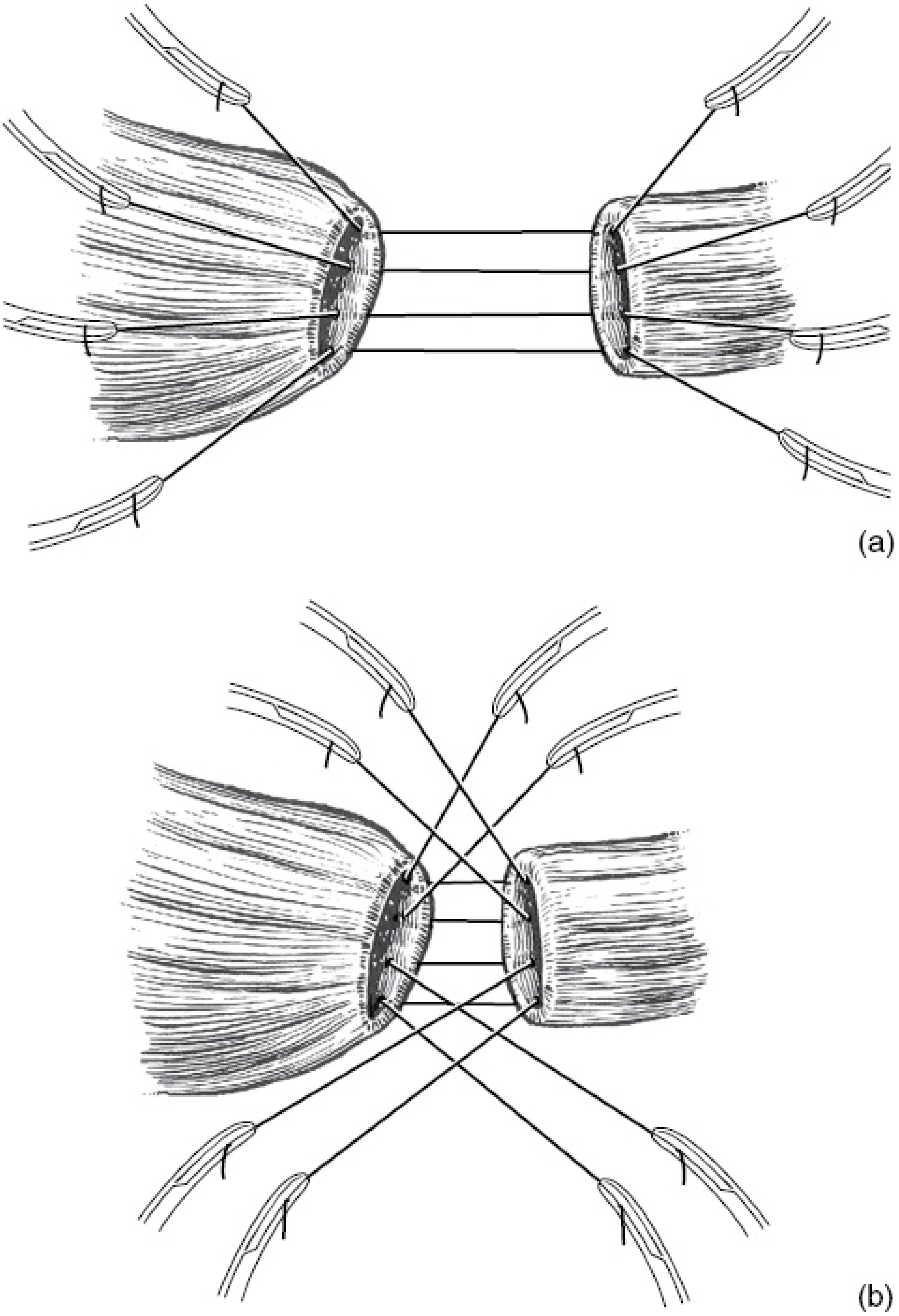
A single layer, end-to-end anastomosis is performed with interrupted absorbable 5/0 or 6/0 sutures (**Figure 9.11**). To start the anastomosis, corner sutures are placed first for alignment. Particular care is taken to ensure that both mucosa and muscle layers are included in each suture. Two or three sutures are then placed with knots on the luminal side to approximate the posterior wall. Unless the two ends of the esophagus are very close together without tension, all posterior wall sutures should be placed before they are tied in sequence.



9.11

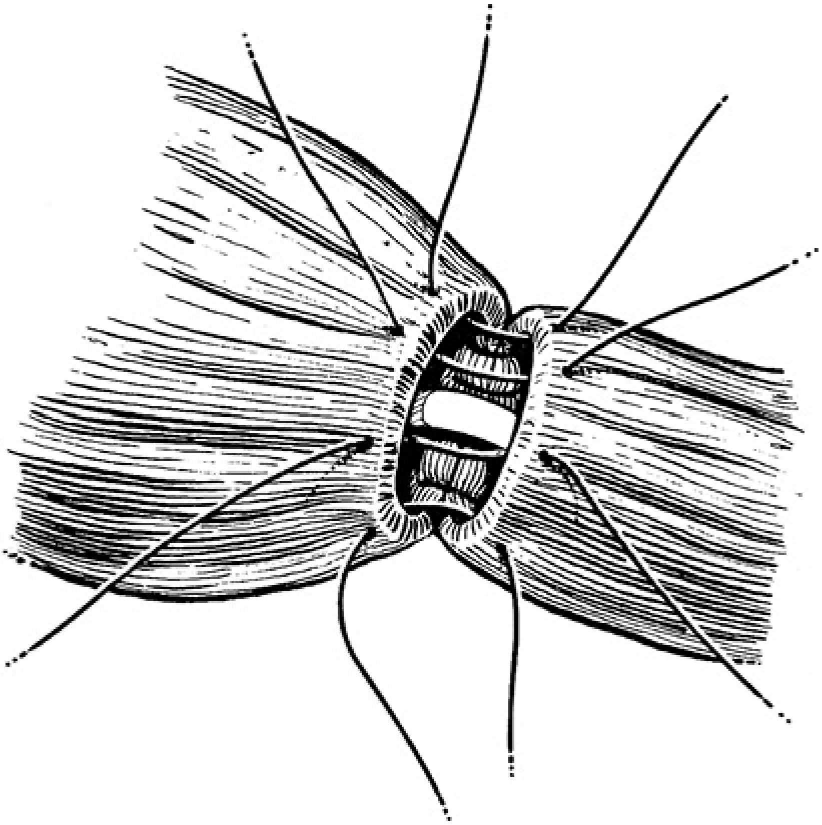
ANASTOMOSIS UNDER TENSION

Once the posterior sutures have been placed (**Figure 9.12a**), the sutures are crossed over and gradual tension is applied, bringing the proximal and distal halves of the posterior esophagus toward each other (**Figure 9.12b**). While maintaining uniform tension on the sutures, each one is tied in turn, thereby distributing the tension on the suture line over a wider area.



9.12a,b

Following completion of the posterior wall of the anastomosis, a 6 Fr or 8 Fr feeding tube is advanced from the upper esophagus, across the anastomosis, and into the stomach to exclude a distal esophageal obstruction (**Figure 9.13**). The anterior wall of the anastomosis is then completed over the tube. The use of a nerve hook avoids the need to use forceps on the open ends of the esophagus when placing the sutures. Once the anastomosis is complete, the tube may be withdrawn since there are data to suggest increased stricture formation with transanastomotic tubes. If the tube is to be left *in situ*, its mobility should be checked to ensure that a suture has not inadvertently passed through or around it. However, recent studies suggest that leaving a transanastomotic tube increases the risk of stricture. Placement of vascularized tissue or mesh between the esophageal and tracheal suture lines has not been shown to have benefit in terms of reducing anastomotic complications.

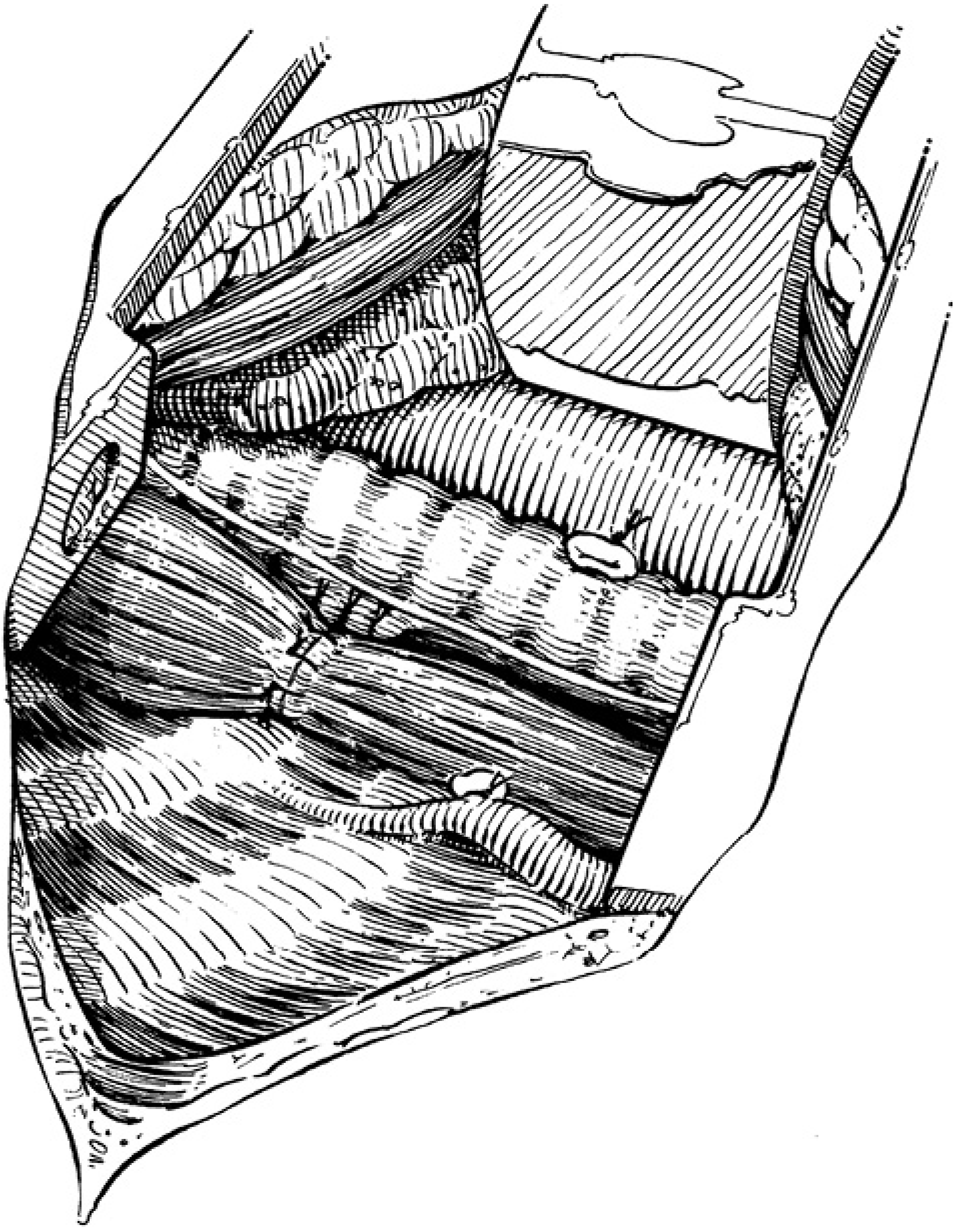


9.13

Although most surgeons leave a retropleural chest drain (12 Fr or 16 Fr), others leave a drain only in specific circumstances (e.g. trauma to the underlying pleura and lung, or occasionally in a very tight anastomosis). If there are concerns regarding chest tube trauma to the anastomosis, the tip of the drain can be sutured to the lateral chest wall away from the anastomotic site with 4/0 chromic catgut.

WOUND CLOSURE

The lung is expanded following the placement of two pericostal 2/0 or 3/0 sutures to reapproximate the rib space (**Figure 9.14**). The muscles and subcutaneous tissues are reapproximated with 4/0 suture and the skin with a subcuticular suture.



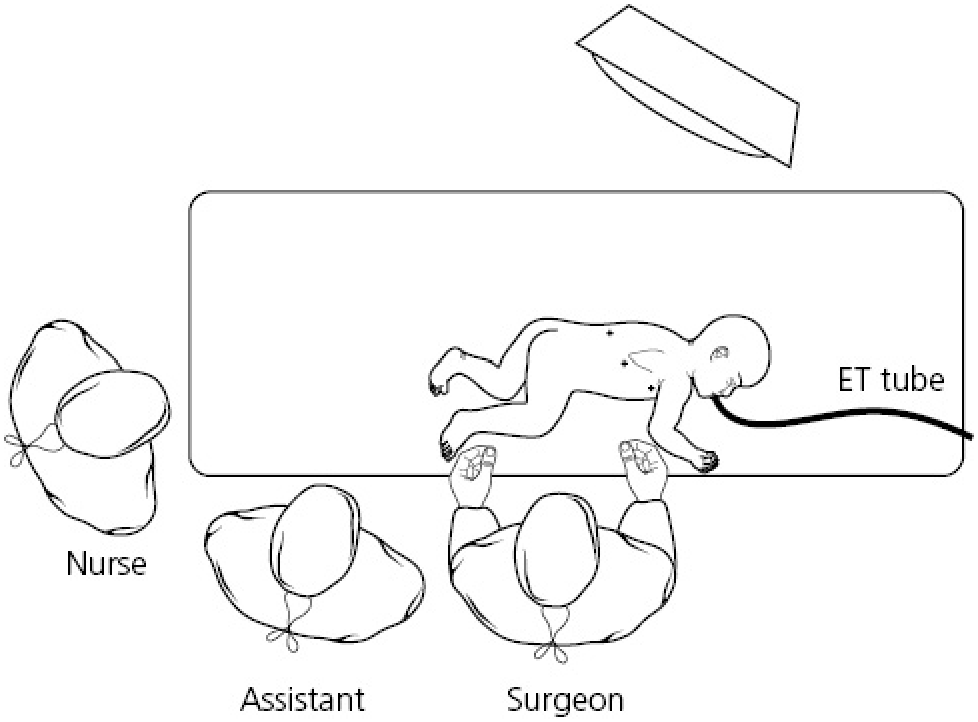
9.14

Minimally invasive (thoracoscopic) repair

The main advantage of thoracoscopic repair of EA with TEF is the reduction in the musculoskeletal sequelae that may develop following thoracotomy in the newborn. Musculoskeletal deformities, including “winged” scapula, asymmetry of the thoracic wall secondary to atrophy of the serratus anterior muscle, and thoracic scoliosis have been reported in 20–40% of children who had a thoracotomy for EA. Mammary maldevelopment has also been reported. Other advantages of thoracoscopic repair may also include better cosmesis and improved anatomic visualization due to significant magnification of the operative field, particularly with regard to the upper pouch dissection. However, the procedure requires advanced endosurgical skills since performing the anastomosis under tension in a small working space can be difficult even for the experienced minimally invasive surgeon. Thoracoscopic repair by those less experienced may result in prolonged operative times and increased risk for anastomotic complications. Neonates less than 2 kg and those with significant cardiac or pulmonary disease may not be good candidates for the thoracoscopic approach.

POSITION

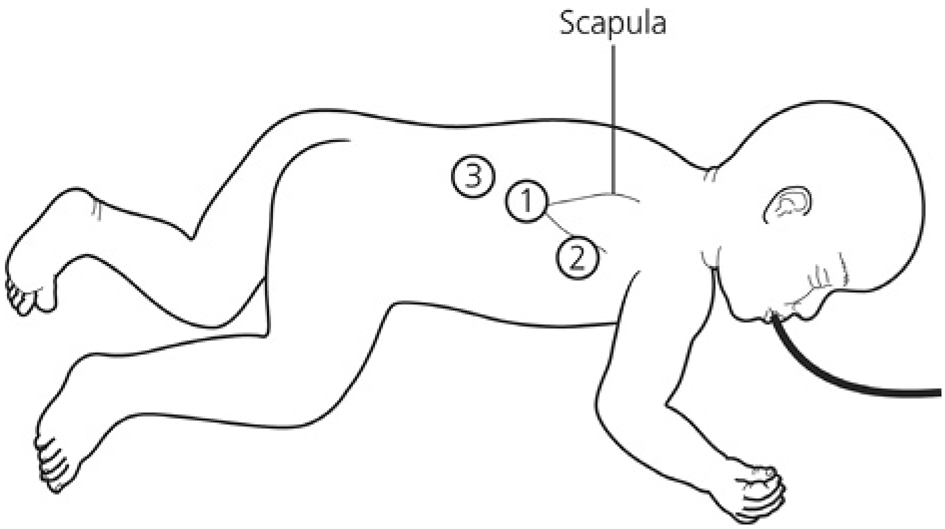
Under general endotracheal anesthesia, the infant is placed in a three-quarter (30-degree) prone position. This will allow the right lung to deflate and fall away from the operation site by natural gravity to expose the operative field without the need for additional ports for retraction. Single lung ventilation is not necessary. The surgeon stands on the left side of the table with the screen opposite at the right upper end of the table. The assistant in most cases is on the surgeon's right, but this may vary depending on the surgeon's and assistant's preferences. The scrub nurse can be at the bottom or other side of the table (**Figure 9.15**).



9.15

ACCESS TO THE CHEST

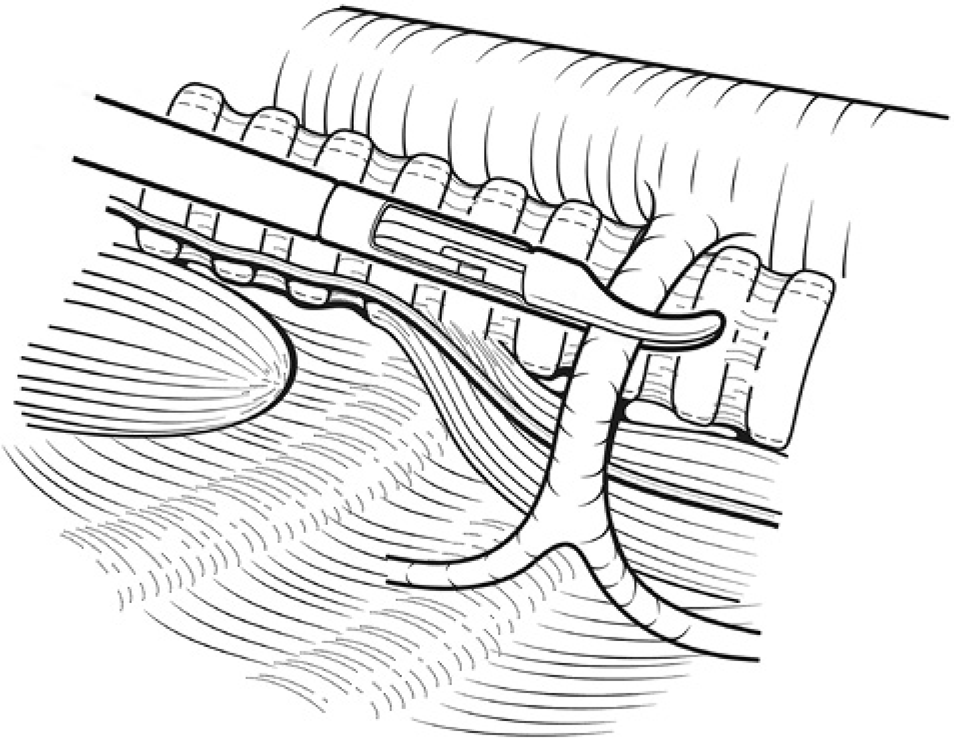
A transpleural approach provides excellent visualization of the posterior mediastinum. The first trocar (4 mm or 5 mm) is inserted just posterior to the tip of the scapula (**Figure 9.16)**. This allows for direct visualization into the posterior mediastinum while looking down on the tips of the instruments. The chest is insufflated with CO_2 to 4–6 mmHg and flow of 1–2 L/min until the lung is collapsed. A 4 mm or 5 mm 30-degree telescope is inserted. A 3 mm port is then placed one or two interspaces below and slightly posterior to the camera port. This is for the use of 3 mm working instruments. Another 3 mm or 5 mm port is inserted in the axilla. In general, this is adequate to visualize the posterior mediastinum. Occasionally, if the lung does not collapse completely, it may be necessary to introduce another 3 mm port posterior to the other two working ports to retract the lung anteriorly with atraumatic forceps.



9.16

It is imperative to monitor oxygen saturations and end tidal CO_2 during the procedure. Many prefer to monitor arterial gases regularly during the operation to minimize respiratory acidosis. More recently, we have been using near-infrared spectroscopy (NIRS) to evaluate cerebral blood and perfusion during the procedure. To avoid this complication, consideration should be given to reducing the insufflation pressure, pausing the operation and allowing the acidosis to correct, and/or converting the operation to the open procedure.

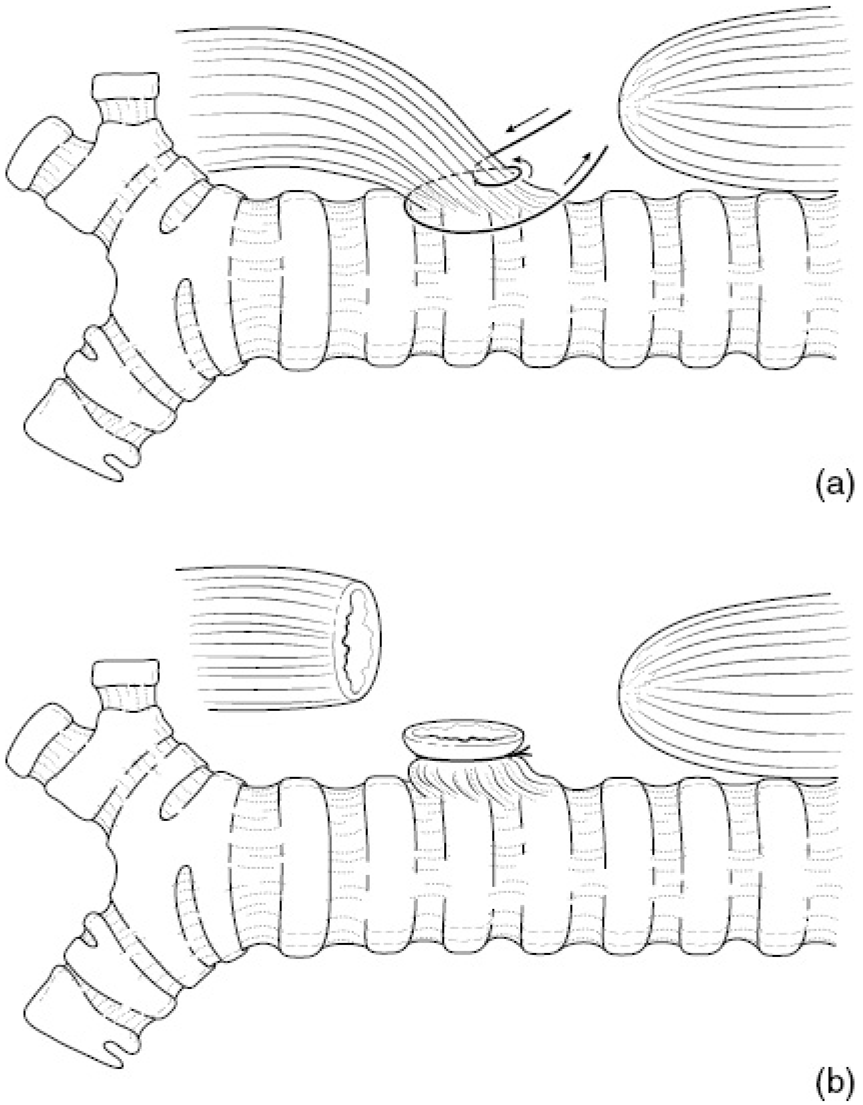
Similar to the open procedure, the azygous vein is identified and transected after coagulation of the vessel using hook diathermy or 3 mm sealer (the authors' preferred technique) (**Figure 9.17**).



9.17

DIVISION OF THE TRACHEOESOPHAGEAL FISTULA

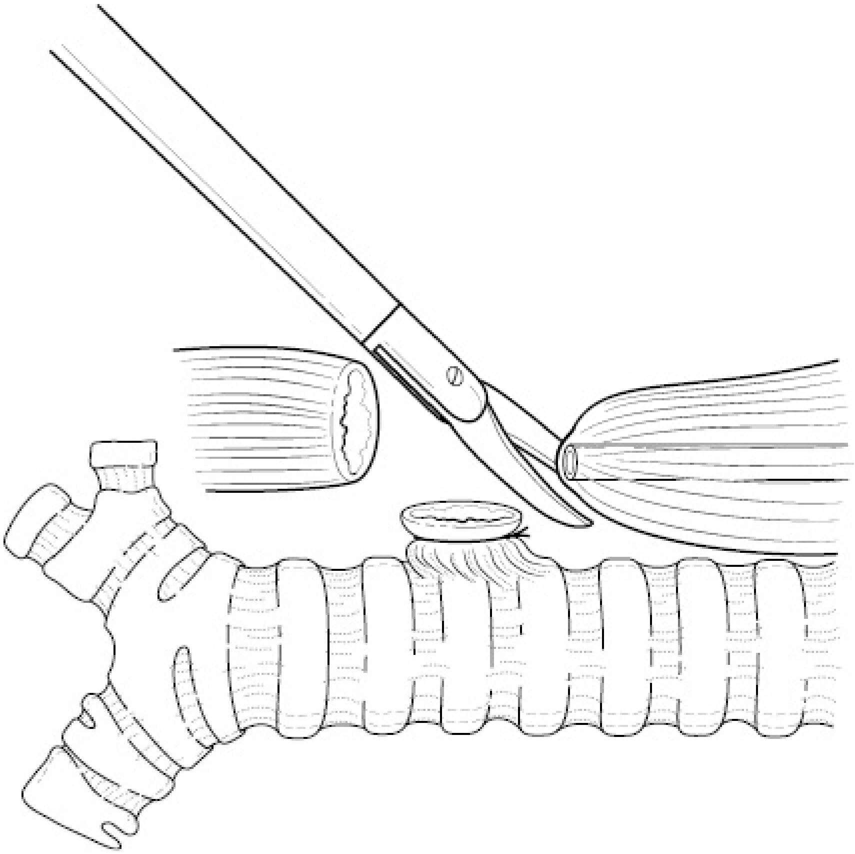
The TEF is visualized during ventilation. Before ligating and dividing the fistula, an atraumatic grasper is used to occlude the fistula and ascertain that the right lung is still ventilating. The fistula is suture ligated close to the posterior tracheal wall using 5 mm endoclips or a 4/0 suture (**Figure 9.18a**). The fistula can now be transected and air leak from the trachea is excluded by Valsalva maneuver (**Figure 9.18b**). Alternatively, this step can be deferred until the time of the anastomosis if there are concerns about retraction of the lower esophagus toward the diaphragm.



9.18a,b

MOBILIZATION OF THE PROXIMAL ESOPHAGEAL POUCH

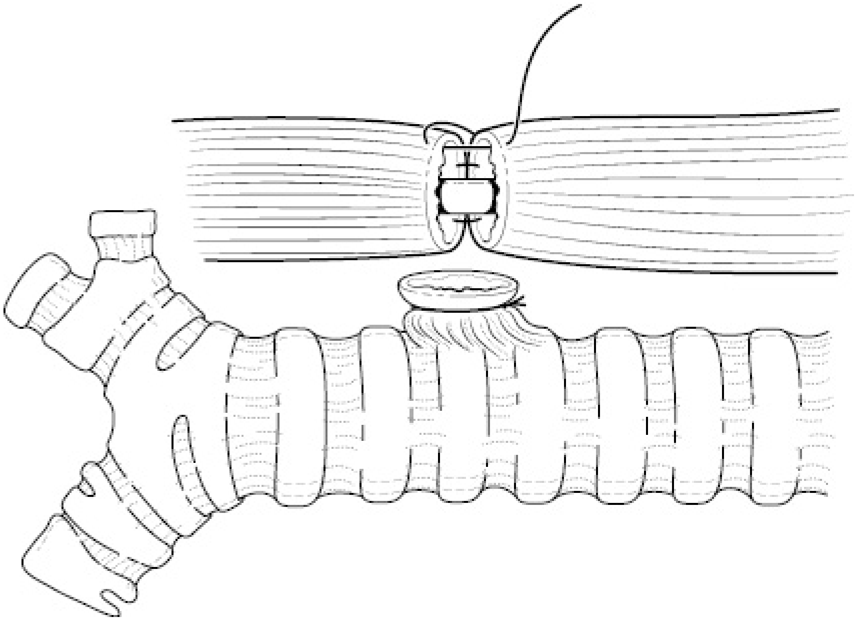
The mediastinal pleura covering the proximal esophageal blind pouch is opened and the pouch mobilized. For guidance, the anesthesiologist can push on the catheter in the proximal esophagus. To help mobilize the upper pouch, in some cases it can be useful to insert a traction suture into the pouch, although in most cases the authors have not found this necessary. The cleavage plane between the proximal esophagus and trachea is developed using scissors to avoid damage to the esophagus and trachea. Hook diathermy is only used when the dissecting is not close to the tracheal wall. Use of the 3 mm sealer (JustRight Surgical, Louisville, CO) to mobilize the upper pouch is also useful and avoids the heat spread caused by the monopolar hook. Dissection can be carried out well into the thoracic inlet to gain considerable length. Once the upper pouch has been sufficiently mobilized, the distal tip of the upper pouch should be resected using dissecting scissors (**Figure 9.19**). This is preferable to just incising the tip and may decrease the incidence of stricture formation.



9.19

ANASTOMOSIS

An end-to-end esophageal anastomosis is performed using interrupted full thickness sutures of absorbable 4/0 or 5/0 suture with intracorporeal knots. Sutures are initially placed in the posterior wall. The anesthesiologist then passes a transanastomotic tube and the anterior part of the anastomosis is completed using the same suture material (**Figure 9.20**). By leaving the end of the suture a little longer, traction can be applied so that the next suture can be placed more easily. As with the open repair, a chest drainage tube is left in place if there is concern for a postoperative tension pneumothorax that may be caused by inadvertent lung injury or if the anastomosis is tenuous. As with the open repair, we no longer routinely leave a transanastomotic tube.



9.20

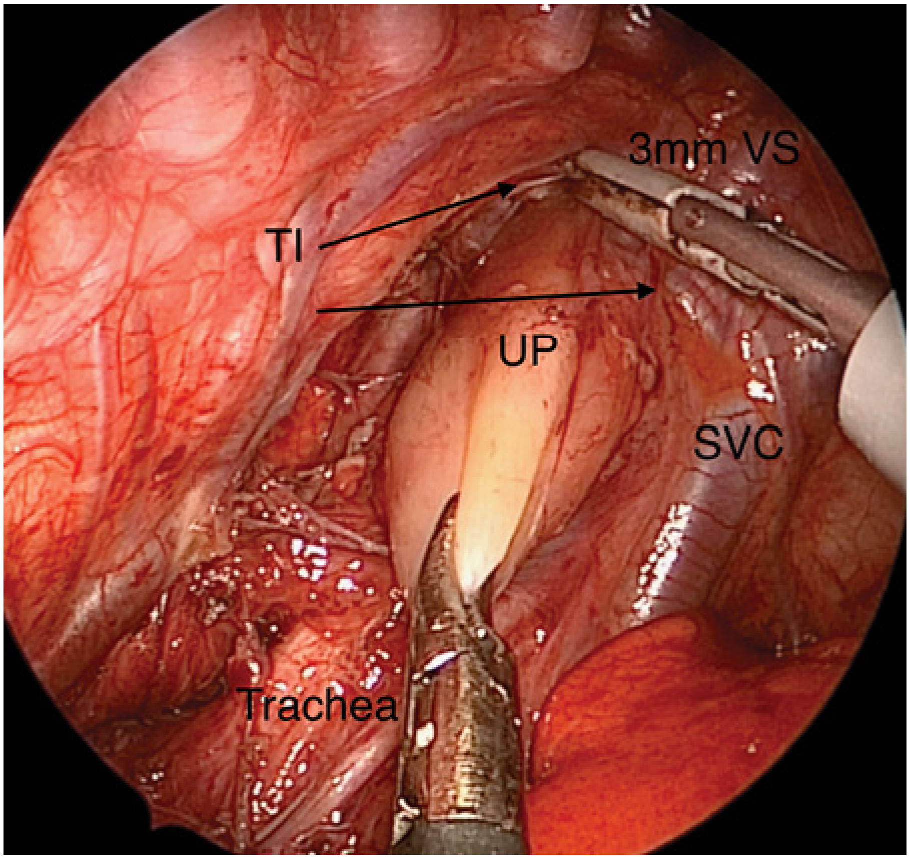
WOUND CLOSURE

The muscle layer of 5 mm port sites is closed with absorbable 4/0 suture. The skin is closed with a subcuticular suture. The postoperative care is as for the open repair.

Long-gap atresia

Long-gap atresia has now been defined by the International Network of Esophageal Atresia (INoEA) as EA with no distal fistula, Gross types A and D. We approach these thoracoscopically as well. Open repair with external traction sutures and the Foker technique will be discussed elsewhere.

The patient usually receives a gastrostomy tube in the first few days of life and then undergoes thoracoscopic exploration at 4–6 weeks of age. We feel little growth of the pouches is realized after this period and the gap may actually become longer as the child's thoracic cavity grows faster than the pouch. The trocar placement is similar to that for a standard TEF except the left-hand port is slightly more inferior and anterior to allow for better dissection of the lower pouch. The key to the success of the operation is extensive mobilization of the upper pouch, which can be accomplished better thoracoscopically because of the direct visualization of the thoracic inlet (**Figure 9.21**). The lower pouch can be mobilized safely down to the diaphragm, but the phrenoesophageal ligament should not be breached as this can increase the risk of a hiatal hernia occuring. Using this technique, we have bridged gaps of up to eight vertebral bodies in a single procedure. If the ends cannot be anastomosed, internal traction sutures can be placed and a repeat thoracoscopy performed in 5–7 days.

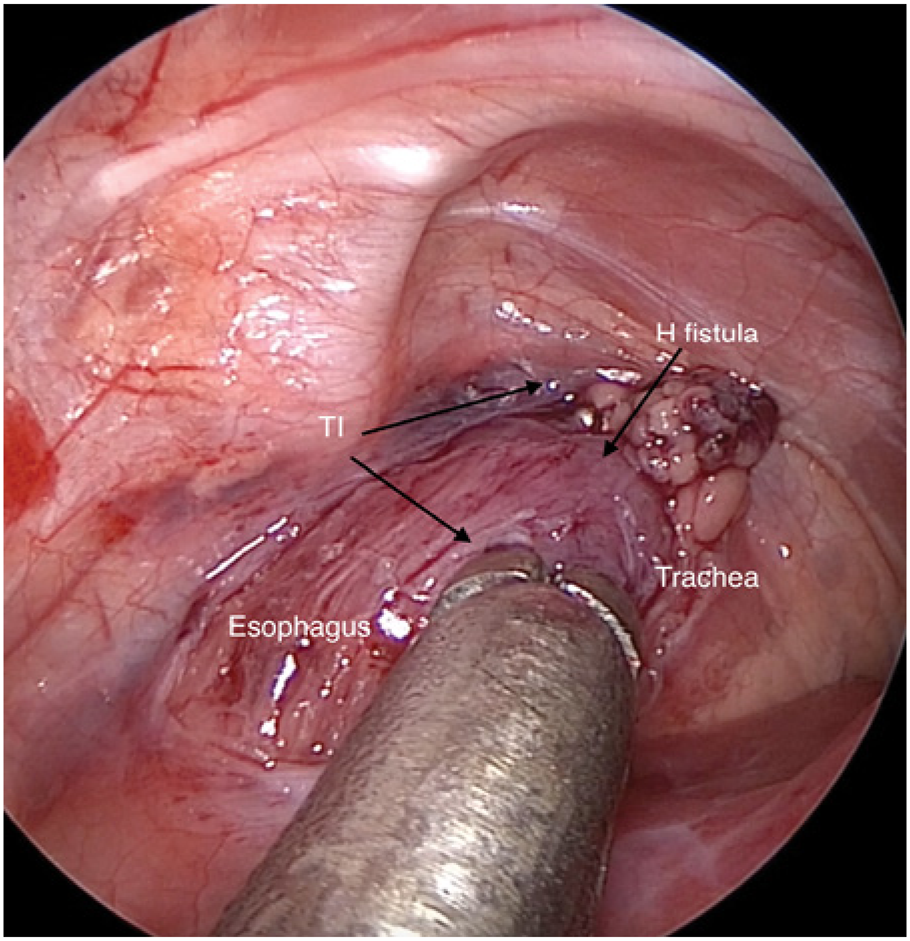


9.21

H-type fistula

The H-type or Gross-type E fistula almost always resides at the thoracic inlet. The traditional approach for an H type fistula has been through a transverse incision just above the clavicle. Dissection is carried down in the anterior cervical triangle and the fistula is found by retracting the sternocleidomastoid muscle and carotid sheath laterally. However, there is a fairly high incidence of injury to the recurrent laryngeal nerve (RLN) as it is hard to visualize from this approach. The fistula is divided sharply and closed with absorbable suture on both sides. In general, it is advisable to interpose tissue between the two cut ends, and often a portion of the sternocleidomastoid can be used.

For the last 10 years we have approached these fistulas thoracoscopically because of the improved visualization of the thoracic inlet (**Figure 9.22**). The fistula can either be suture ligated or more recently we have used the 5 mm stapler to divide the fistula. The hope is that improved visualization will decrease the incidence of RLN injury.



9.22

POSTOPERATIVE CARE

Regardless of the operative approach, the infant is nursed in the neonatal intensive care unit. If the esophageal anastomosis has been performed under extreme tension, some recommend paralysis and mechanical ventilation for several days. Intravenous fluids are administered and antibiotic prophylaxis is continued. A postoperative esophagram is performed approximately 4–5 days after the procedure to check the integrity of the anastomosis. If there is no anastomotic leak, oral feeding can be gradually introduced. Regular chest physiotherapy, with nasopharyngeal suction, as required, is carried out to avoid respiratory infection.

OUTCOME

Published single-center and multicenter studies of EA show favorable results for thoracoscopic repair compared to neonates undergoing repair by thoracotomy. To date, there have been no randomized controlled trials comparing the thoracoscopic and open approaches.

COMPLICATIONS

- Early
  - Anastomotic leakage
  - Anastomotic stricture
  - Vocal cord paralysis
- Late
  - Recurrence of fistula
  - Gastroesophageal reflux
  - Tracheomalacia
  - Esophageal dysmotility

Anastomotic leaks occur in 15–25% of cases, but major disruption is rare (<2%). Minor leaks are generally detected during the esophagram and can be managed non-operatively with antibiotics and chest tube drainage. Major leaks often present in the first 48 hours postoperatively with a tension pneumothorax and sepsis. In these patients, consideration should be given to re-exploration of the anastomosis with the intention of repairing the leak.

Anastomotic strictures develop in 15–45% of cases, most responding to serial dilations. Risk factors include anastomotic tension, anastomotic leakage, and gastroesophageal reflux. Recurrent TEFs develop in approximately 5% of cases and present with choking or cyanotic attacks with feeding or with recurrent episodes of pneumonia.

FURTHER READING

Holcomb GW 3rd, Rothenberg SS, Bax KM, et al. Thoracoscopic repair of esophageal atresia and tracheoesophageal fistula: A multi-institutional analysis. *Ann Surg* 2005; 242: 422–8.

Holland AJ, Ron O, Pierro A, et al. Surgical outcomes of esophageal atresia without fistula for 24 years at a single institution. *J Pediatr Surg* 2009; 44: 1928–32.

Kunisaki SM, Bruch SW, Hirschl RB, et al. The diagnosis of fetal esophageal atresia and its implications on perinatal outcome. *Pediatr Surg Int* 2014; 30: 971–7.

Lal D, Miyano G, Juang D, et al. Current patterns of practice and technique in the repair of esophageal atresia and tracheoesophageal fistua: An IPEG survey. *J Laparoendosc Adv Surg Tech A* 2013; 23: 635–8.

Lal DR, Gadepalli SK, Downard CD, et al. Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula. *J Pediatr Surg* 2017; 52: 1245–51.

Lopez PJ, Keys C, Pierro A, et al. Oesophageal atresia: Improved outcome in high-risk groups? *J Pediatr Surg* 2006; 41: 331–4.

Rothenberg SS. Thoracoscopic repair of esophageal atresia and tracheo-esophageal fistula. *Semin Pediatr Surg* 2005; 14: 2–7.

Rothenberg SS. Thoracoscopic management of non-type C esophageal atresia and tracheoeophageal atresia. *J Pediatr Surg* 2018; 53(1): 121–5.

Rothenberg SS, Flake AW. Experience with thoracoscopic repair of long gap esophageal atresia in neonates. *J Laparoendosc Adv Surg Tech A* 2015; 25: 932–5.

Spitz L, Kiely EM, Morecroft JA, et al. Oesophageal atresia: At-risk groups for the 1990s. *J Pediatr Surg* 1994; 29: 723–5.

van der Zee DC, Bax KN. Thoracoscopic treatment of esophageal atresia with distal fistula and of tracheomalacia. *Semin Pediatr Surg* 2007; 16: 224–30.

10

Esophageal replacement with colon

NAZIHA KHEN-DUNLOP, CARMEN CAPITO, CHRISTOPHE CHARDOT, AND YANN RÉVILLON

HISTORY

The first description of the anatomic details for esophageal replacement with colon is credited to Vuillet and Kelling in 1911. Von Hacker was the first to perform the procedure 3 years later. For many years, the colon was the primary substitute for a damaged or atretic esophagus. Many of the pitfalls using the transposed colon in adults stem from problems with vascular supply to the graft. Even if these problems are less commonly seen in the pediatric population, it has given way to newer techniques, using gastric or jejunal interposition.

Long-gap atresia and caustic injury are the two main indications for graft interposition in the pediatric population while other indications, such as esophageal tumors, button batteries ingestion, diffuse candidiasis, intractable achalasia, and epidermolysis bullosa, remain exceptional.

PRINCIPLES AND JUSTIFICATION

Even though preservation of the patient's own esophagus should always be attempted, persisting with attempts to retain the native esophagus at all costs could be detrimental to the patient. Motility may not be normal and not allow normal oral feeding. In reality, the choice of the surgical approach seems to depend more on local preference and experience than on objective data. Globally, stomach interposition seems to be preferred for long-gap esophageal atresia repairs while colon graft are proposed for both esophageal atresia and caustic burns with a division of two-thirds gastric interposition and one-third colon interposition.

The incidence of caustic injuries has dramatically decreased in developed countries with the introduction of childproof containers, and the majority of these patients now come from developing countries. Persistent strictures after 12 months of repeated dilatations or temporary stenting indicate the need for replacement.

We do not think there is an ideal approach yet. Colon has an excellent mobile vascular supply via the marginal artery that always provides adequate length to reach up into the chest or the neck. It has also the appropriate caliber for esophageal anastomosis. We believe that its main advantage compared to gastric interposition is the absence of Barrett's esophagus, because of absent acid production in the conduit and the resistance of colonic mucosa to acid reflux from the stomach. However, there are two main problems with colon interposition, namely an initial precarious blood supply and long-term redundancy.

The minimal age for colon replacement is still controversial. Some authors advocate waiting until children start walking because of the higher mortality rates in the neonatal period. However, colonic interposition has been performed from 6 months of age by more experienced teams with good results. In younger age groups, the small size of colonic vessels may lead to an increased risk of graft vascular compromise and, in the older child, the blood supply remains precarious compared to gastric blood supply. Meticulous selection and manipulation of the transposed colon remains vital to avoid graft necrosis.

The normal esophagus contracts with forceful peristaltic contractions in response to initiation of a swallow; by contrast the normal colon contracts in a more complex pattern resulting from distension of the colon and is dependent on both hormonal and neurogenic factors. Thus, the colon interposed into the esophageal bed may need a larger volume of distension before emptying. Animal studies of isolated segments of colon showed more rapid emptying in isoperistaltic versus antiperistaltic loops, but in both cases colon redundancy can progressively occur.

PREOPERATIVE

Preparation

Most children with caustic burns come from developing countries and they may have a poor nutritional state and preoperative feeding programs are necessary. During the initial siting of the gastrostomy, the middle colon artery could be ligated in order to increase the vascularity of its left branch by increasing the marginal vessel flow.

Prior to esophageal replacement, the child is admitted and clear liquids are given for 24 hours preoperatively. Polyethylene glycol electrolyte solution is given at a rate of 25–40 mL/kg per hour until stools are clear.

Anesthesia

The procedure is carried out under general endotracheal anesthesia. An epidural catheter can facilitate the intraoperative anesthesia and postoperative pain control.

OPERATION

Principles

- Esophagectomy, as extensive as possible, has to be performed in patients where damage has been caused by caustic burns, because of the possible risk of malignancy in the remnant.
- Both retrosternal or mediastinal esophagoplasty are possible, but the posterior way is shorter without kinking and is preferred when possible.

The right transverse or left colon can be mobilized and brought up to the neck in most children without compromise of the blood supply to the graft.

Most surgeons use the transverse colon based on the marginal artery of Drummond. The graft is placed in the isoperistaltic direction, without graft tension to prevent distal ischemia, but not too long to limit secondary colon redundancy.

Prevention of graft ischemia should be kept in mind at each step of the surgical procedure.

- Take care when selecting the main pedicle of the graft and during the clamping tests of its auxiliary blood supply.
- It is important when "storing" the graft during the esophagectomy, and elevating it via the prepancreatic retrogastric route. Avoid a very narrow pedicle and check for absence of volvulus.
- When moving the graft in its mediastinal route, we use a lubricated bag (echography probe bag or similar) passed through the mediastinal route to gently pull the graft to the neck inside the bag, reducing the risk of stripping the pedicle.
- The esophagocolon is made without tension or twist, with interrupted stitches.

There is a high incidence of postoperative proximal anastomotic leaks. Even if the higher part of the esophagus seems to be intact and retainable, a cervical anastomosis avoids the potential disaster of an intrathoracic leak and acute mediastinal sepsis.

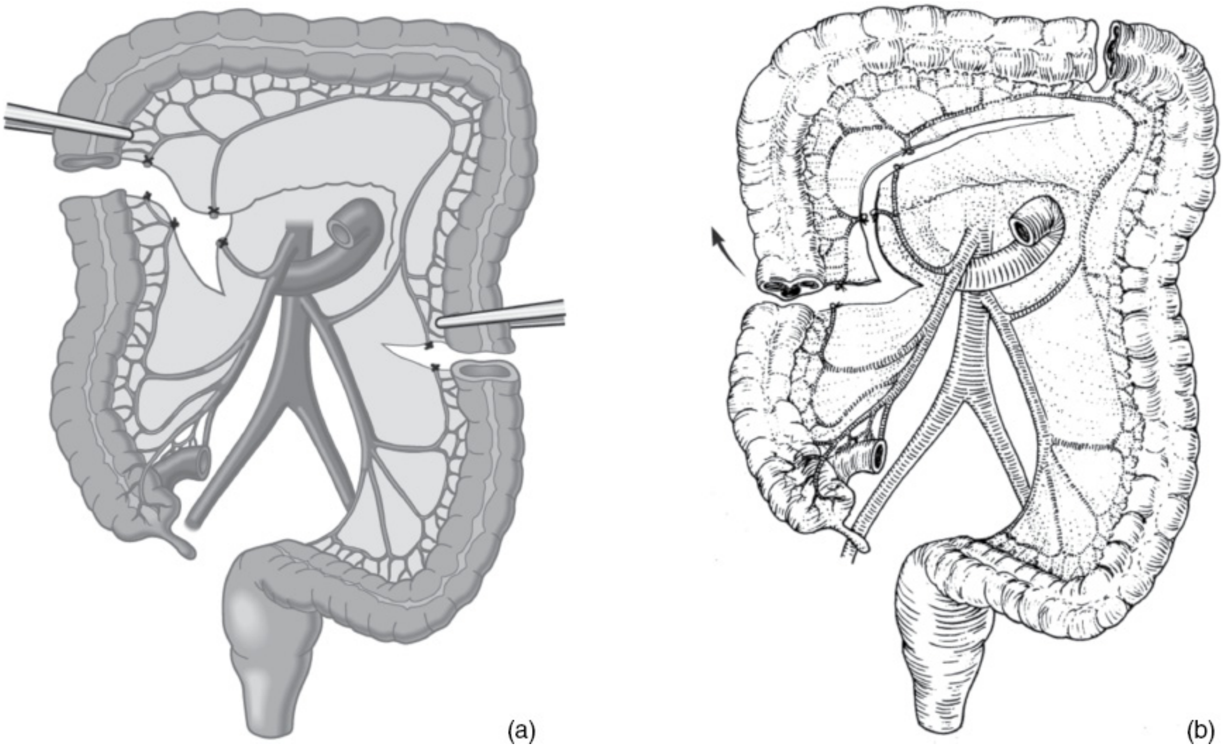
Position of patient

The patient is placed in a 45-degree left lateral decubitus position with the right arm prepped into the operative field. The table is rolled laterally to afford greater exposure for the abdominal portion of the procedure.

Abdominal incision

The procedure is started abdominally because it confirms the technical availability of a well-vascularized colon graft and may avoid a thoracotomy if the esophagectomy is possible via a transhiatal dissection.

A midline incision is made and the colon is examined. Vascular supply to the colon dictates the segments to be transposed into the chest. The blood supply of the right colon is very inconstant and the typical anatomical description is observed in less than a quarter of cases. Once the pedicle is chosen, mostly transverse colon, small vascular clamps are placed on the middle colic artery, if not ligated previously, and the marginal artery to evaluate the graft perfusion before dividing the vessels. At the same time, intestinal clamps are placed at the two extremities of the graft to stop the transmural vascularization (**Figure 10.1a and b**). The perfusion of the proposed graft by the remaining blood supply is assessed after 10 minutes and, if adequate, the clamped vessels are divided centrally, preserving collaterals in the marginal artery system of the colon segment (**Figure 10.1a and b**).



10.1a,b

A posterior mediastinal blunt dissection is performed. An umbilical tape is passed from the colonic mesentery through the diaphragmatic hiatus to the level of the proximal esophageal segment in the neck. This tape helps determine the length of colon necessary for replacement. Accurate measurement of the distance between the site of the upper and lower anastomosis is necessary to avoid graft tension and resection of excessive tissue may be necessary to prevent secondary colon redundancy. The colon is then divided at an appropriate length.

Esophagectomy

In caustic burns, esophagectomy is most often performed via a blind transhiatal approach. This procedure is probably one of the most difficult steps of the surgery because of the adhesions, particularly at the level of the aortic arch and the left bronchus. Laparoscopic assistance, through a transhiatal approach, has been proposed for mediastinal dissection and esophagectomy, based on lower morbidity than conventional blunt dissection and with a higher rate of the total esophagus removal (perhaps reaching 80%). Video surgery is also becoming more useful because it reduces operative trauma on nerves and the trachea and ensures precise hemostasis.

Thoracic incision

A posterolateral thoracotomy incision is made in the sixth intercostal space in cases of mid-esophagus stricture. If there has been no previous thoracotomy, an extrapleural approach can be made. A subperiostal resection of the sixth rib with incision into the extrapleural space through the posterior periosteum may facilitate the approach. A transpleural approach is made in the case of previous thoracotomy. The lung is retracted anteromedially and the proximal esophagus identified with the aid of the manipulation of a nasoesophageal tube. The proximal esophagus is mobilized to the thoracic inlet and followed by the esophagectomy. The distal esophageal segment is mobilized to the esophageal hiatus. In children with minimal scarring of the esophageal bed, transhiatal dissection and total esophagectomy without thoracotomy is possible.

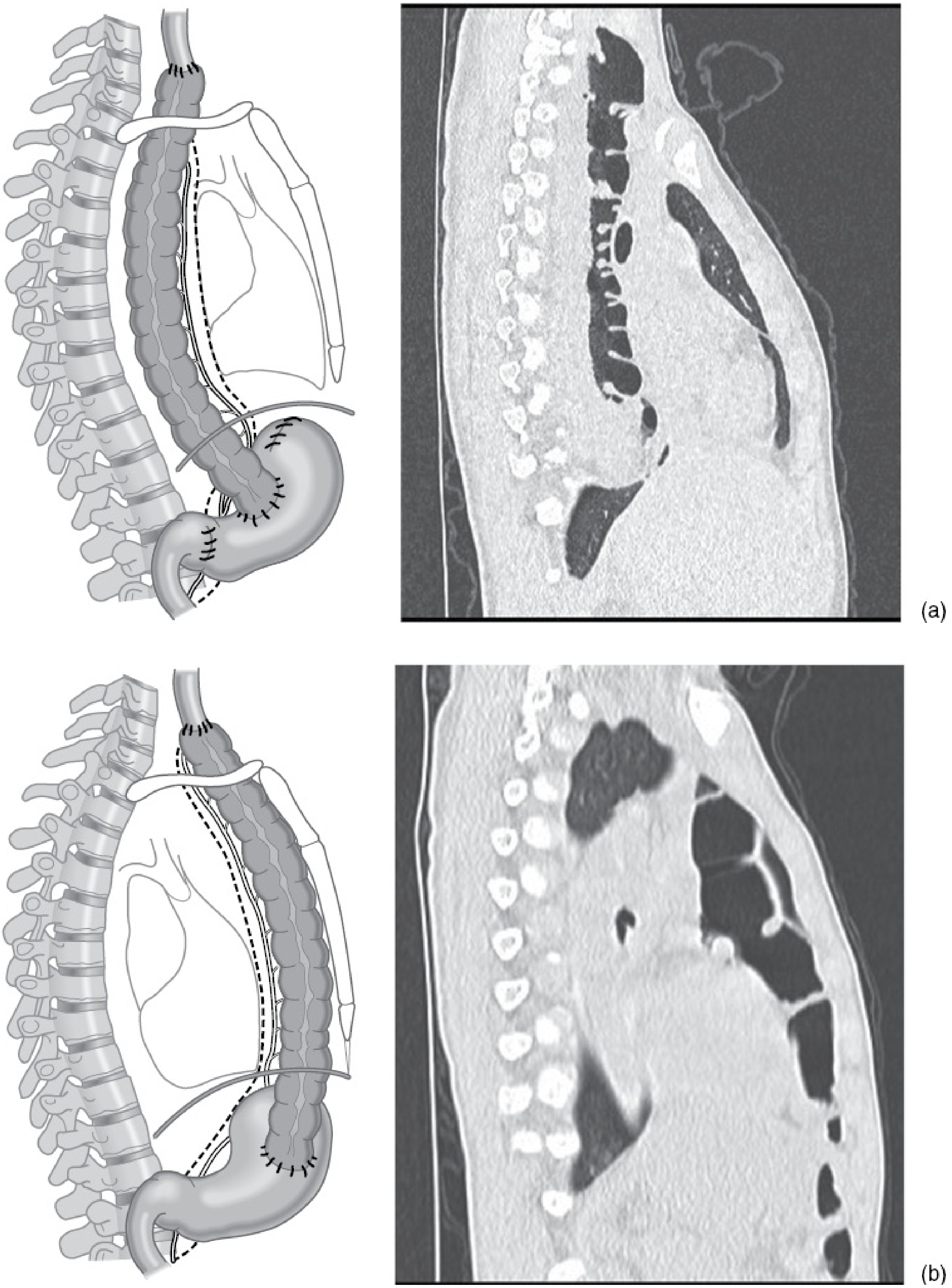
Cervical incision

Minimal dissection of the esophagus is carried out in patients with previous esophagostomy to prevent injury to the recurrent laryngeal nerves and to minimize esophageal ischemia. If not, an oblique or transverse right neck incision is made. The neck vessels are retracted and the proximal esophagus is mobilized within the thoracic inlet.

In those cases with the most severe caustic burns, there may be damage to the pharynx, epiglottis, and pyriform sinuses. Thorough evaluation of the mobility of the vocal cords is essential and, if intact, an entire resection with a proximal anastomosis at the level of the arytenoids can be possible. This very complex procedure needs active collaboration with of ear, nose, and throat (ENT) surgeons.

Colon interposition

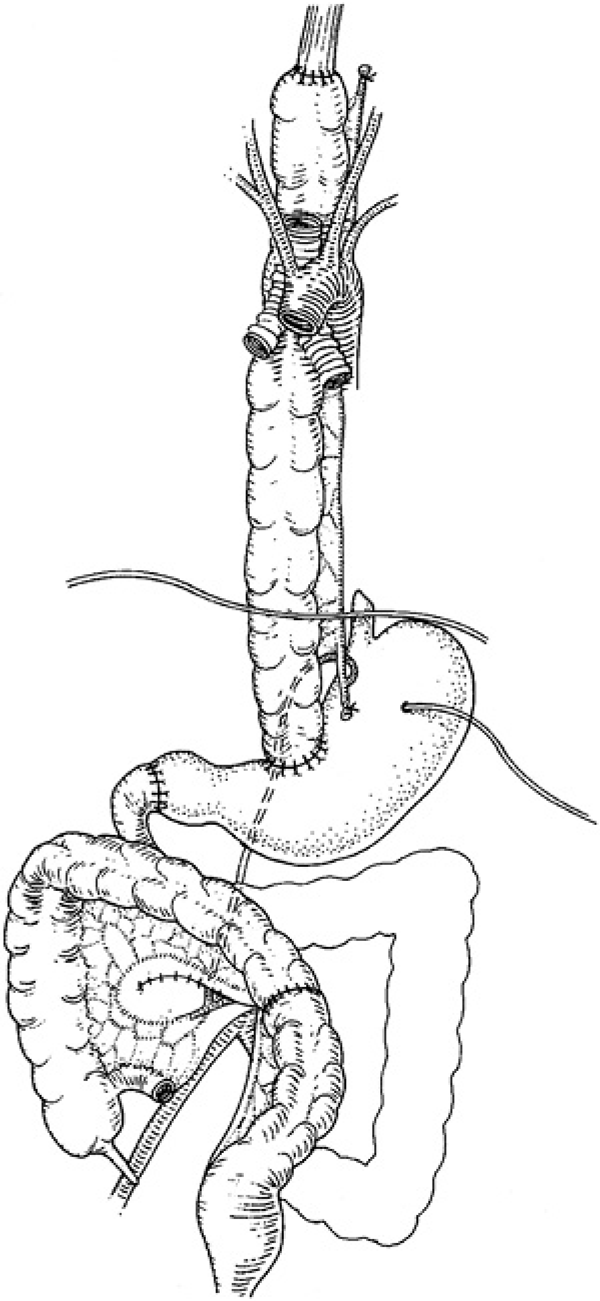
Once the colon graft is mobilized, the distal esophageal stump is resected, if possible at the stomach with a linear stapling device to avoid reflux in the stump. In some cases, a normal gastroesophageal junction may be preserved, anastomosing the distal esophagus to the colon. Normally, the graft is pulled through the chest, into the posterior mediastinum (**Figure 10.2a**). Scarring in the native esophageal bed may preclude this and a substernal route may be used (**Figure 10.2b**). If this is chosen, removal of the sternoclavicular joint or a part of the manubrium may be necessary to enlarge the space and avoid a kink at the thoracic inlet. Numerous devices have been used, from plastic laparoscopy camera cable covering to a large Penrose drain, to help facilitate the passage of the graft.


(a)
(b)

10.2a,b

Anastomosis

The proximal anastomosis between the esophagus and colon is created in the neck with a single layer of absorbable sutures and the colon is fixed to the neck muscles. The gastrocolic anastomosis is performed at the anterior gastric wall, to limit gastrocolonic reflux, and the anastomosis is partially wrapped with the stomach (**Figure 10.3**). Pyloroplasty is usually performed and (temporary) gastrostomy is left for gastric decompression and transition to oral feeds.



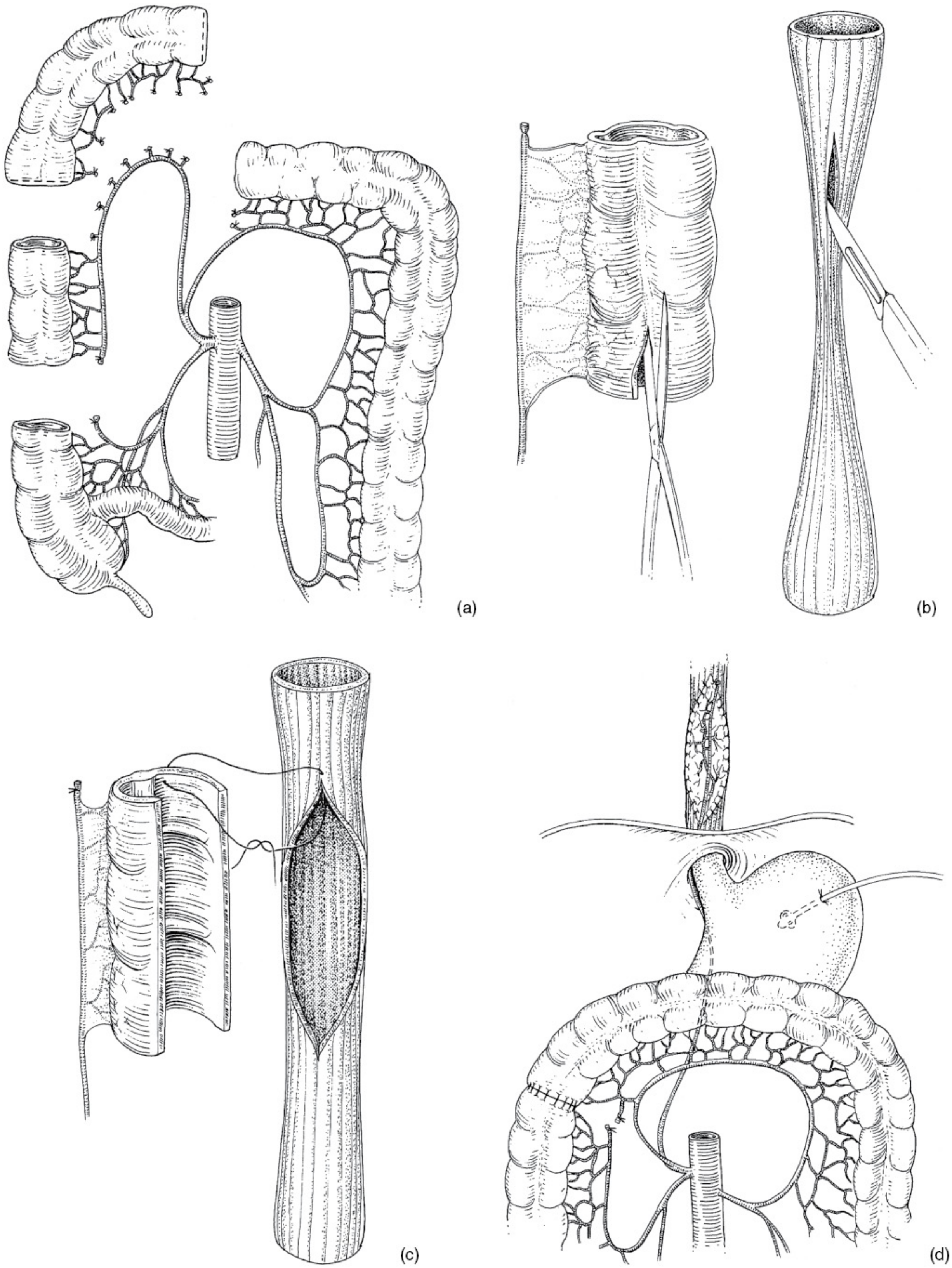
10.3

The colocolic anastomosis finishes the procedure, anterior to the vascular pedicle. A nasocolonic tube is also placed in the interposed colon for postoperative graft decompression to prevent distension.

Colonic patch esophagoplasty

Although not a common operation nowadays, colonic patch esophagoplasty can sometimes be helpful as an alternative to esophageal replacement.

A thoracic approach is made, exposing the area of stricture. The colon is then mobilized and measured via an abdominal incision, and a satisfactory pedicle graft is created, as if performing an esophageal replacement procedure. Those segments based on the right colic artery are preferred (**Figure 10.4a**).


(a)
(b)
(c)
(d)

10.4a–d

The esophageal stricture is then opened and the length of stricture to be patched is determined. A segment of colon equal to the length of the esophageal defect is measured (**Figure 10.4b**). The segment of colon distal to the patch segment but adjacent to the mobilized vascular pedicle is removed. The marginal artery is carefully preserved by dividing vessels close to the wall of the segment of bowel to be discarded. The colon patch segment is opened along its antimesenteric border (**Figure 10.4c**). The patch is then passed behind the stomach through the esophageal hiatus (**Figure 10.4d**). If the patch is too large, a pseudodiverticulum may develop. A single layer of interrupted or continuous absorbable suture is used to secure the patch in place.

POSTOPERATIVE CARE

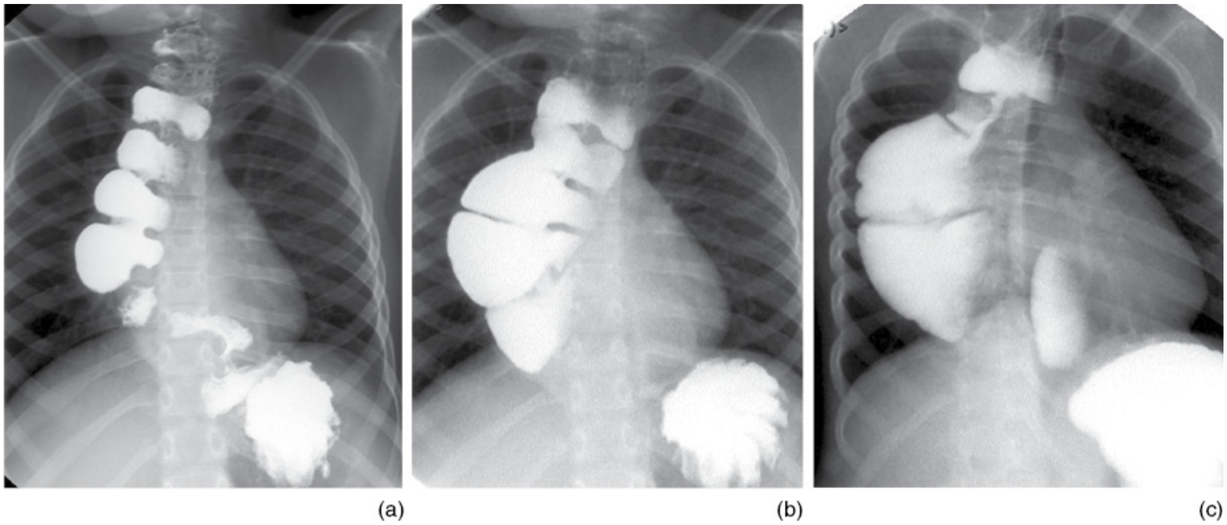
The intracolonic tube is removed on the third day postoperatively if no graft distension is seen on chest radiograph. A contrast swallow is performed on the seventh postoperative day. If there is no leakage, oral feeding may be started and the chest drains may be removed.

OUTCOME

Esophageal replacement is a demanding surgical procedure with important judgments made on the optimal procedure. The outcomes of colon interposition compared to gastric transposition are still debated. Both have an equivalent mortality risk but subjective appreciation of the outcome seems to vary considerably.

Dumping symptoms may occur, but these are less common than after gastric transposition. Post-cibal discomfort, vagal reactions, or seizures after intake of food suggest hypoglycemia. An apparent disadvantage is the need for three anastomoses and hence a threefold increase in the risk of leakage. However, our experience shows that this complication mainly and almost exclusively concerns the cervical anastomosis. Distal ischemia because of a precarious vascular supply may, of course, be one cause, but caustic burns particularly may have an effect on the cervical esophagus, increasing the risk of anastomotic failure. The rate of theses cervical leaks is high, about 30–40%, but they resolve spontaneously in most cases. Formation of secondary strictures can then be observed and treated with endoscopic dilatations. In rare cases, resection of the stricture is, however, necessary, but after a complete re-evaluation of the esophageal integrity.

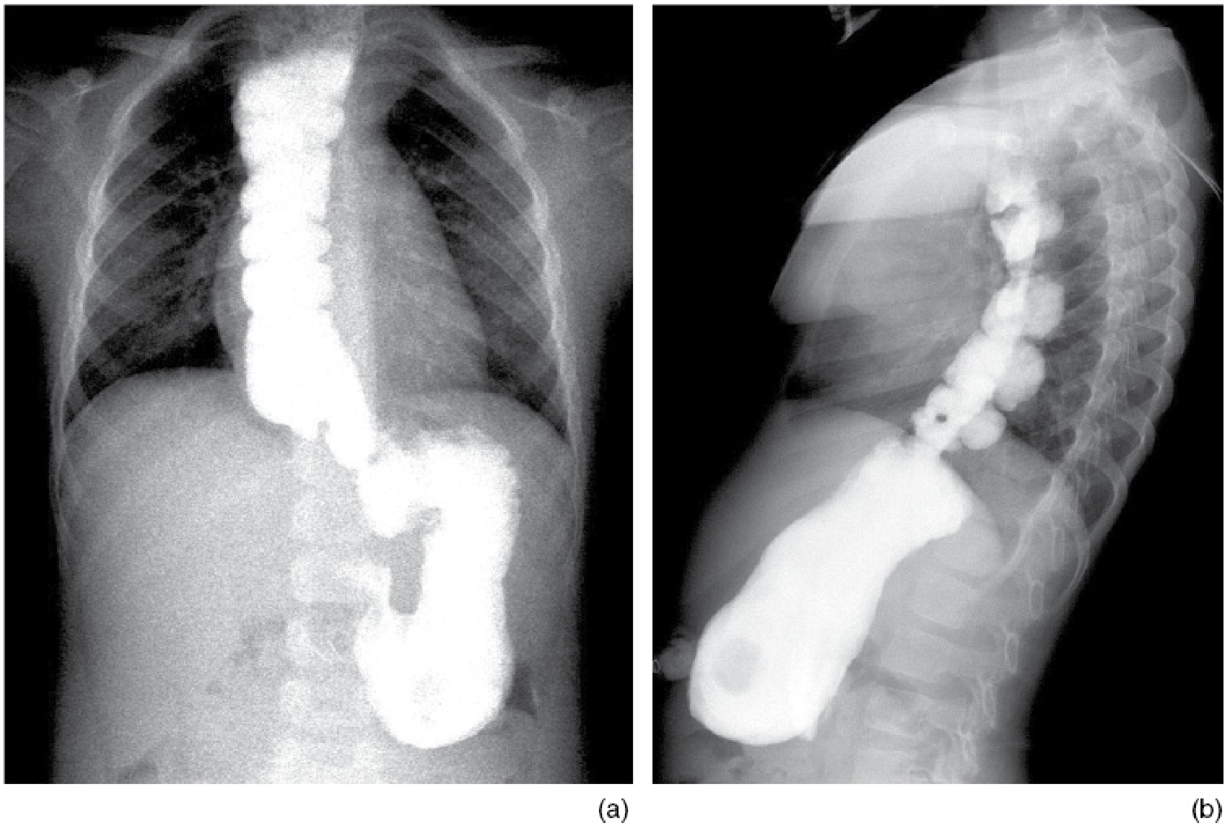
The main problem of esophageal replacement with colon is redundancy. The colon graft segment does not establish peristaltic contractions coordinated with swallowing as in the native esophagus, thus emptying is primarily by gravity and solid transit is slow. Consequently, the colon interposition becomes redundant over time, leading to obstruction, dysphagia, and possible aspiration (**Figure 10.5a–c**). In symptomatic cases, resection of the redundant part is necessary. Results are excellent but this surgery is very complex. Because of local inflammation secondary to the chronic stasis and the fibrosis of the ancient burn scars, mobilization of the colon graft is difficult and a careful and permanent control of the vascular supply is necessary



10.5a–c

Postoperative complications are observed in about 50% of cases, with respiratory problems and local infections common, but mortality remains low at 1–5%. Late complications are also more frequent, with scoliosis reported in 25% of cases and reflux in 40%.

Full oral feeding is usually achieved within 6 months. Most patients who have undergone esophageal replacement with colon lead an otherwise normal life and the quality of life is deemed “acceptable.” Despite a high morbidity, colon interposition remains a proven, satisfactory, durable replacement for the esophagus in children that allows normal swallowing (**Figure 10.6a and b**). Because of the risks of late strictures and excessive tortuosity of the neoesophagus, long-term follow-up is necessary.



10.6a,b

The “colon patch” theoretically avoids many of the functional disadvantages of the segmental colon interposition, and problems related to redundancy and difficulty with emptying are not seen. Endoscopic evaluations of patients with colon patch have also shown re-epithelialization and scar regression. However, it includes a long intrathoracic suture line, thereby increasing the potential for perioperative leakage and secondary pseudodiverticulum. Moreover, this approach is only possible for very limited strictures with the certainty of normal esophagus around.

FURTHER READING

Burgos L, Barrena S, Andrés AM, et al. Colonic interposition for esophageal replacement in children remains a good choice: 33-year median follow-up of 65 patients. *J Pediatr Surg* 2010; 45: 341–5.

Esteves E, Sousa-Filho HB, Watanabe S, et al. Laparoscopically assisted esophagectomy and colon interposition for esophageal replacement in children: Preliminary results of a novel technique. *J Pediatr Surg* 2010; 45: 1053–60.

Kunisaki SM, Coran AG. Esophageal replacement. *Semin Pediatr Surg* 2017; 26: 105–15.

Lima M, Destro F, Cantone N, et al. Long-term follow-up after esophageal replacement in children: 45-year single-center experience. *J Pediatr Surg* 2015; 50: 1457–61.

Reinberg O. Esophageal replacements in children. *Ann N Y Acad Sci* 2016; 1381: 104–12.

11

Esophageal replacement with transposition of the stomach

PAOLO DE COPPI

INTRODUCTION

Gastric transposition has the advantage of involving only one well-vascularized anastomosis that is associated with a low incidence of leakage.

HISTORY

In 1922, Kummell described the technique of gastric transposition via the mediastinal route in two patients, both of whom died. In 1945, Richard Sweet from Boston, MA, recorded 12 esophageal resections with esophagogastric anastomosis above the aortic arch. Six children, with good long-term results in four, were then reported by John Atwell in Southampton, UK, in 1980. The technique was then popularized by Lewis Spitz and Edward Kiely from Great Ormond Street Hospital in London, who described the successful outcome of 34 infants and children in 1987.

Notably, gastric transposition is currently the procedure of choice for esophageal replacement in adults with esophageal carcinoma.

OPERATION

The procedure is performed transhiatally via the posterior mediastinum without having to resort to a thoracotomy. During the past decade, we have introduced the laparoscopic-assisted gastric transposition, which has now become our routine procedure. If the posterior mediastinum is severely scarred from previous surgery, the left pleural or retrosternal route may be preferred. A thoracoabdominal approach is rarely required.

The importance of sham feeds in infants with long-gap esophageal atresia who have undergone a cervical esophagostomy in simplifying the initiation of oral nutrition following the interposition should not be underestimated. Temporary jejunostomy should always be considered in any case of poor feeding abilities.

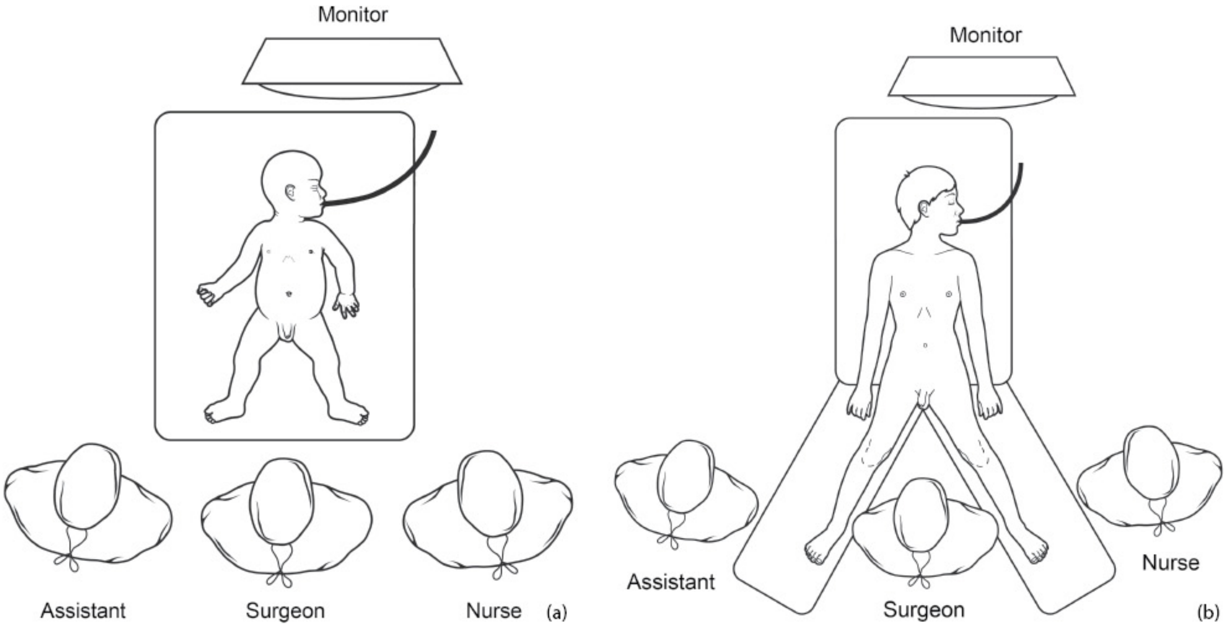
Initial gastrostomy

The initial feeding gastrostomy should ideally have been sited on the anterior surface of the body of the stomach, well away from the greater curvature, in order to preserve the vascular arcades of the gastroepiploic vessels.

POSITION OF PATIENT (LAPAROSCOPIC AND OPEN)

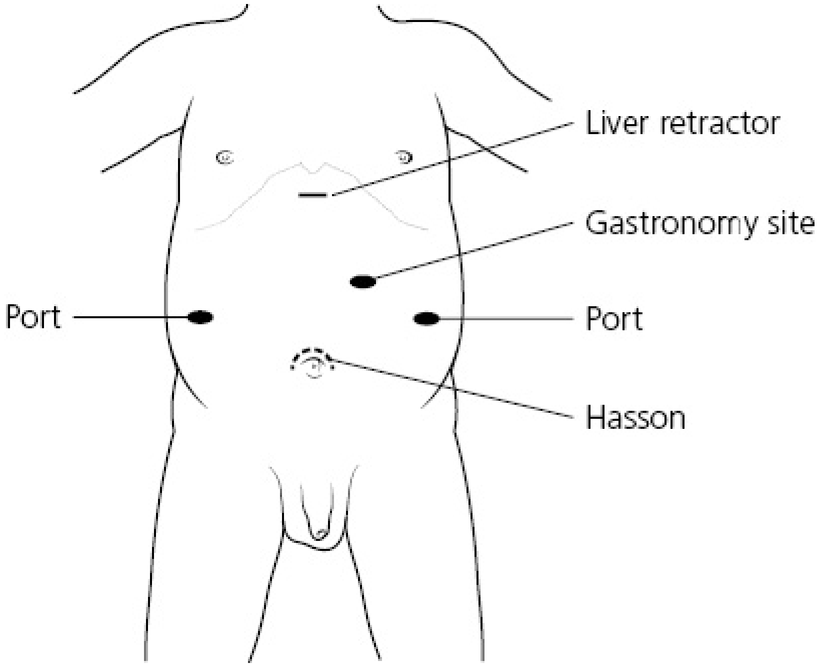
An upper abdominal midline incision or a left upper abdominal transverse muscle-cutting incision is used in the open approach.

The child is positioned at the foot of the operating table according to the floor plan and the video monitor is placed at the head of the table (**Figure 11.1a**). The table should be tilted head-up about 20–30 degrees. In older children, the legs are abducted using a special split table and the operator works between the legs at the bottom of the table; the position of the video monitor is as above (**Figure 11.1b**).



11.1a,b

The laparoscopic approach is performed with three ports. A primary Hasson port for the telescope (5 mm) is usually placed above the umbilicus with two instrument ports in the right and left upper quadrants of the abdomen (**Figure 11.2**). It is important to insert these ports well laterally so that a wide angle is created that facilitates intracorporeal suturing. The port on the right upper quadrant can be 3 mm or 5 mm depending on the size of the patient. The authors prefer a 5 mm disposable port in the left upper quadrant since it allows the introduction of needles for suturing and accommodates both 3 mm and 5 mm laparoscopic instruments. In the midline of the epigastrium, a small incision is made for the introduction of a Nathanson liver retractor without insertion of a port. The liver retractor size is decided on the basis of the size of the liver and the child. The liver retractor is kept in position with the help of a fast clamp.



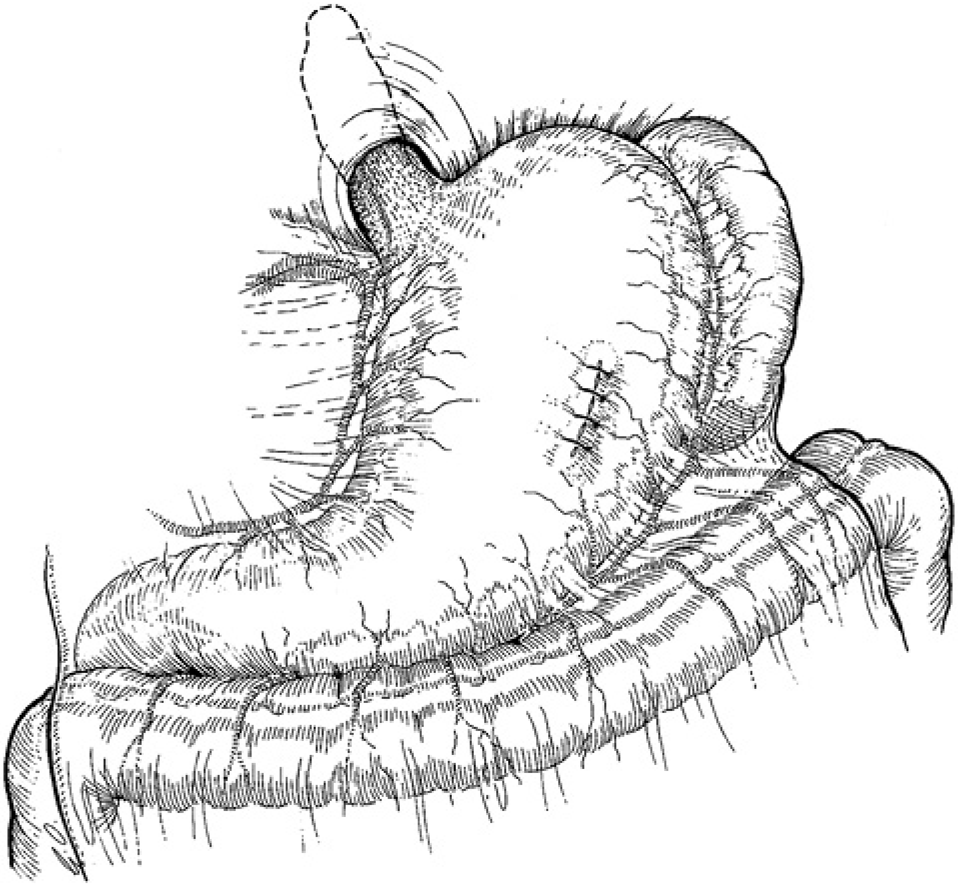
11.2

The gastrostomy is carefully mobilized from the anterior abdominal wall and the defect in the stomach closed in two layers with interrupted 4/0 Vicryl□ acid sutures.

MOBILIZING THE STOMACH

Adhesions between the stomach and the left lobe of the liver are released, taking care not to damage any of the major blood vessels.

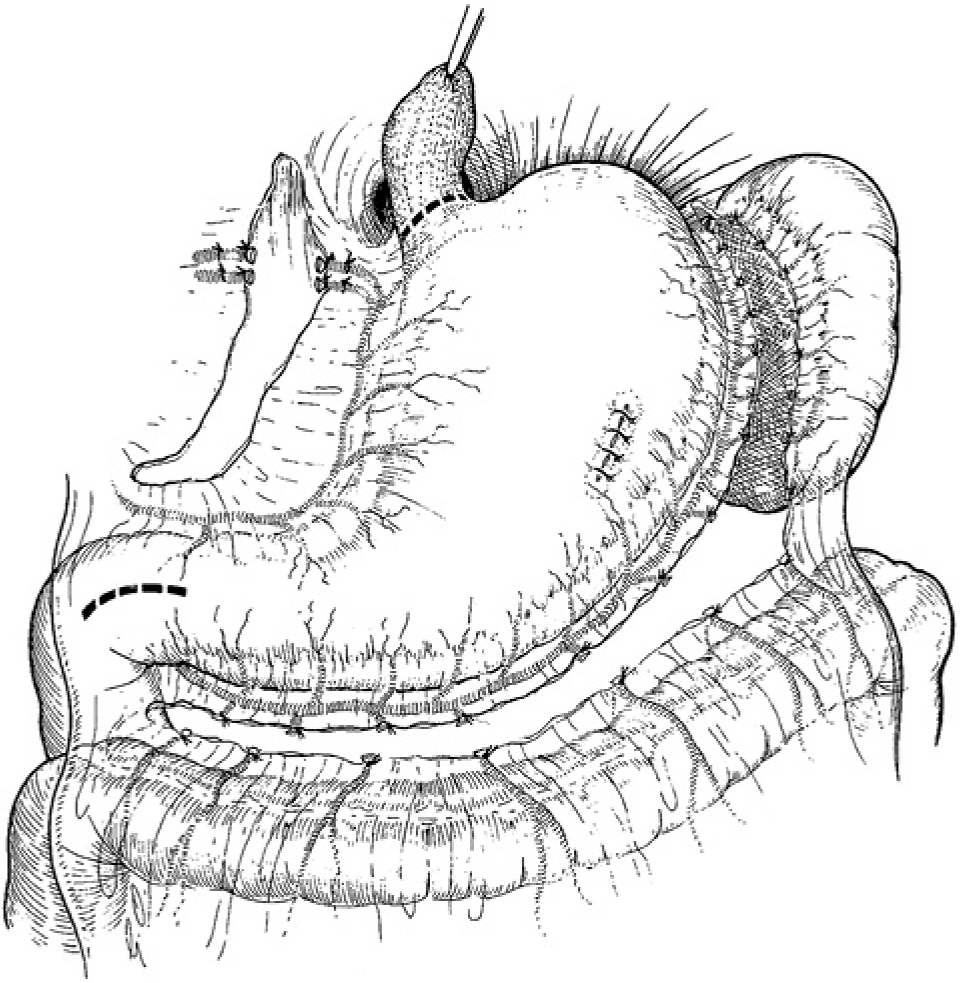
The greater curvature of the stomach is mobilized using a vessel sealing device to divide vessels in the gastrocolic omentum and the short gastric vessels (**Figure 11.3**). These vessels should be divided well away from the stomach wall in order to preserve the vascular arcades of the right gastroepiploic vessels.



11.3

The lesser curvature of the stomach is freed by dividing the lesser omentum from the pylorus to the diaphragmatic hiatus. The right gastric artery is carefully identified and preserved, while the left gastric vessels are divided close to the stomach. The lower esophagus is exposed by dividing the phrenoesophageal membrane, and the margins of the esophageal hiatus in the diaphragm are defined.

In the laparoscopic approach, the Nathanson liver retractor can be used to retract the liver upwards, allowing mobilization of the stomach. When a failed open or laparoscopic fundoplication and/or a gastrostomy has been performed previously, adhesions will be present between the stomach and the liver. These adhesions are carefully divided using hook diathermy and the liver retractor is moved in the desired position. Mobilization of the greater and lesser curve of the stomach is performed following the same principles described in the open technique (**Figure 11.4**). The right gastric artery is identified and preserved and the left gastric vessels are divided using a Harmonic scalpel™.



11.4

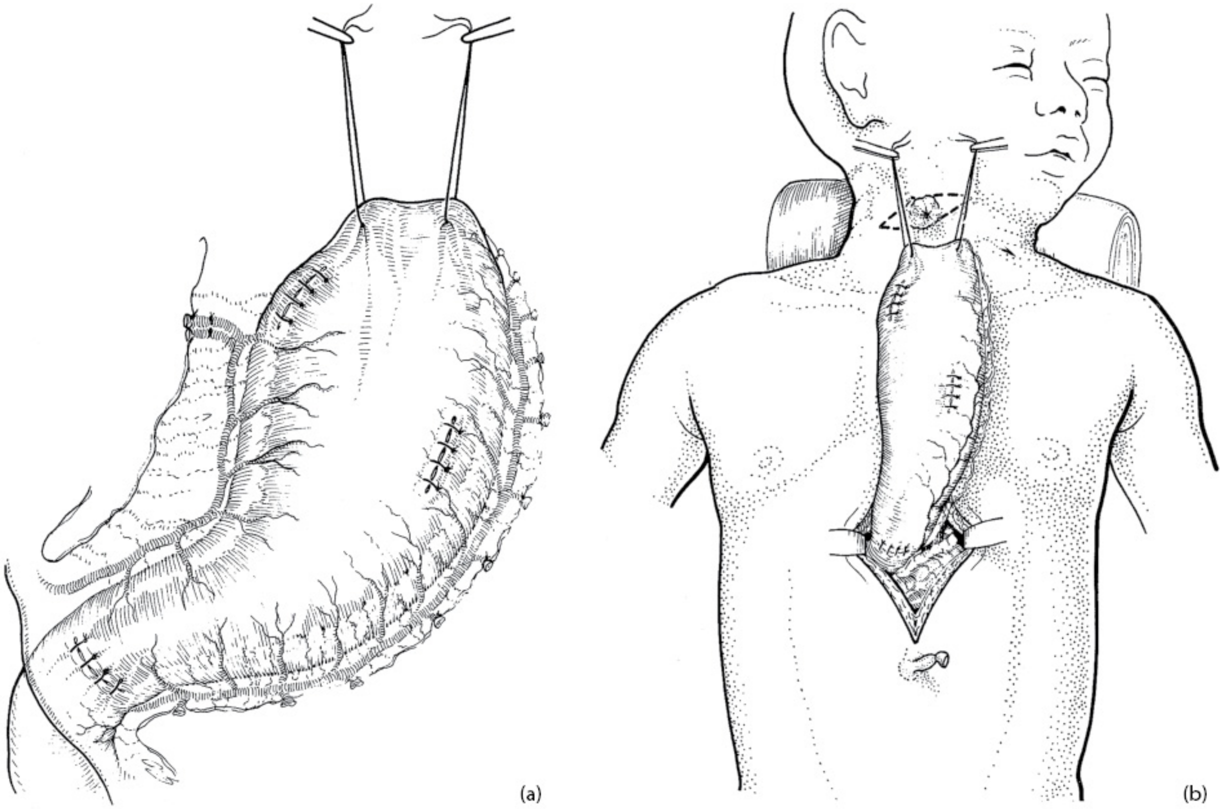
RESECTION OF THE DISTAL ESOPHAGUS

Switching to a zero-degree camera in the laparoscopic dissection helps the dissection of the intrathoracic inevitably short, blind-ending lower esophageal stump by a combination of blunt and sharp dissection through the diaphragmatic hiatus. The anterior and posterior vagal nerves are divided during this part of the procedure. The body and fundus of the stomach are now free from all attachments and can be delivered into the wound.

The esophagus is transected at the gastroesophageal junction and the defect closed in two layers with 4/0 Vicryl™ acid sutures or using staples. This procedure can be done through the opening of the gastrostomy site when the laparoscopic approach is used to shorten the procedure. Rarely, the second part of the duodenum may be Kocherized to obtain maximum mobility of the pylorus.

PREPARING FOR GASTROESOPHAGEAL ANASTOMOSIS

The highest part of the fundus of the stomach is identified and stay sutures of different material are inserted to the left and the right of the area selected for the anastomosis (**Figure 11.5a**). These sutures help to avoid torsion of the stomach occurring as it is pulled up through the posterior mediastinum into the neck (**Figure 11.5b**).



11.5a,b

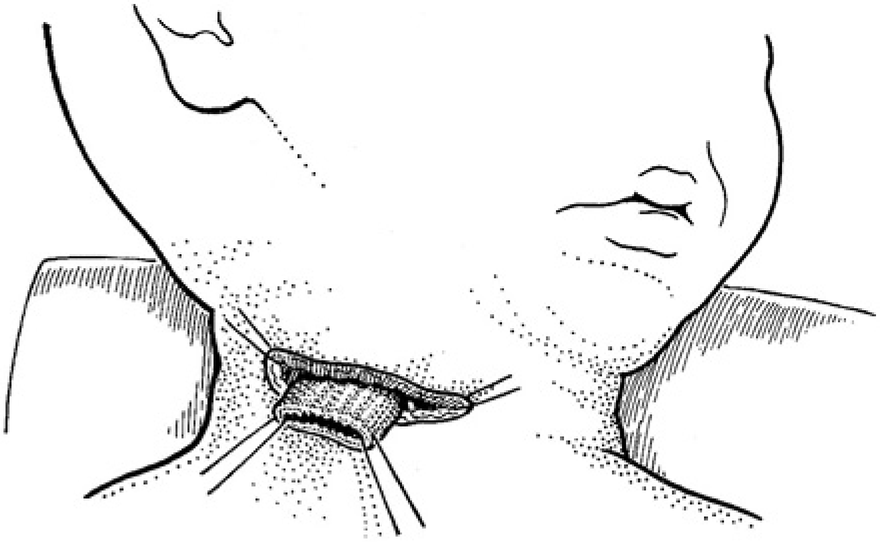
PYLOROPLASTY

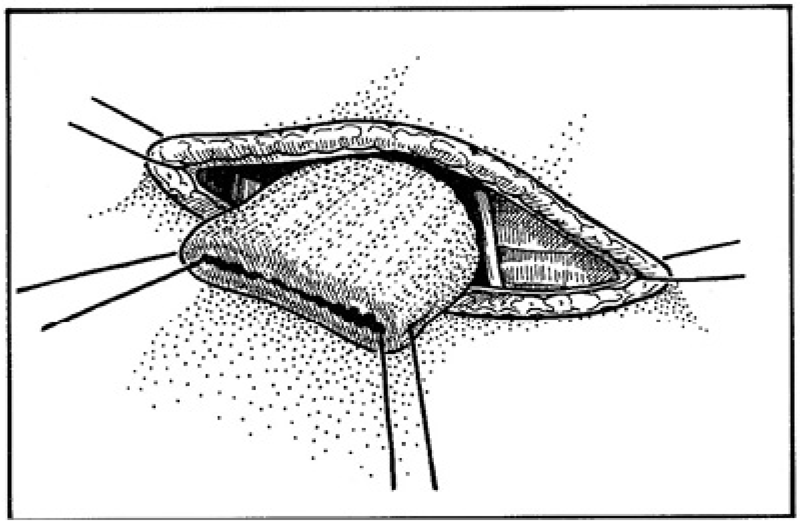
All patients should receive a Heinecke–Mikulicz pyloroplasty, with the transverse incision being closed horizontally with interrupted fine Vicryl™ sutures.

The stomach is now ready for its transposition into the neck.

MOBILIZATION OF THE CERVICAL ESOPHAGUS

Attention is now turned to the neck, where the previously constructed cervical esophagostomy (preferably performed on the right side) is mobilized via a 3–4 cm transverse incision, taking care not to damage the muscular coat of the esophagus. The recurrent laryngeal nerve coursing upwards on the posterolateral surface of the trachea is identified and preserved. It is important to mobilize at least 1.5–2 cm full-thickness esophagus to allow a satisfactory anastomosis to take place (**Figure 11.6**).

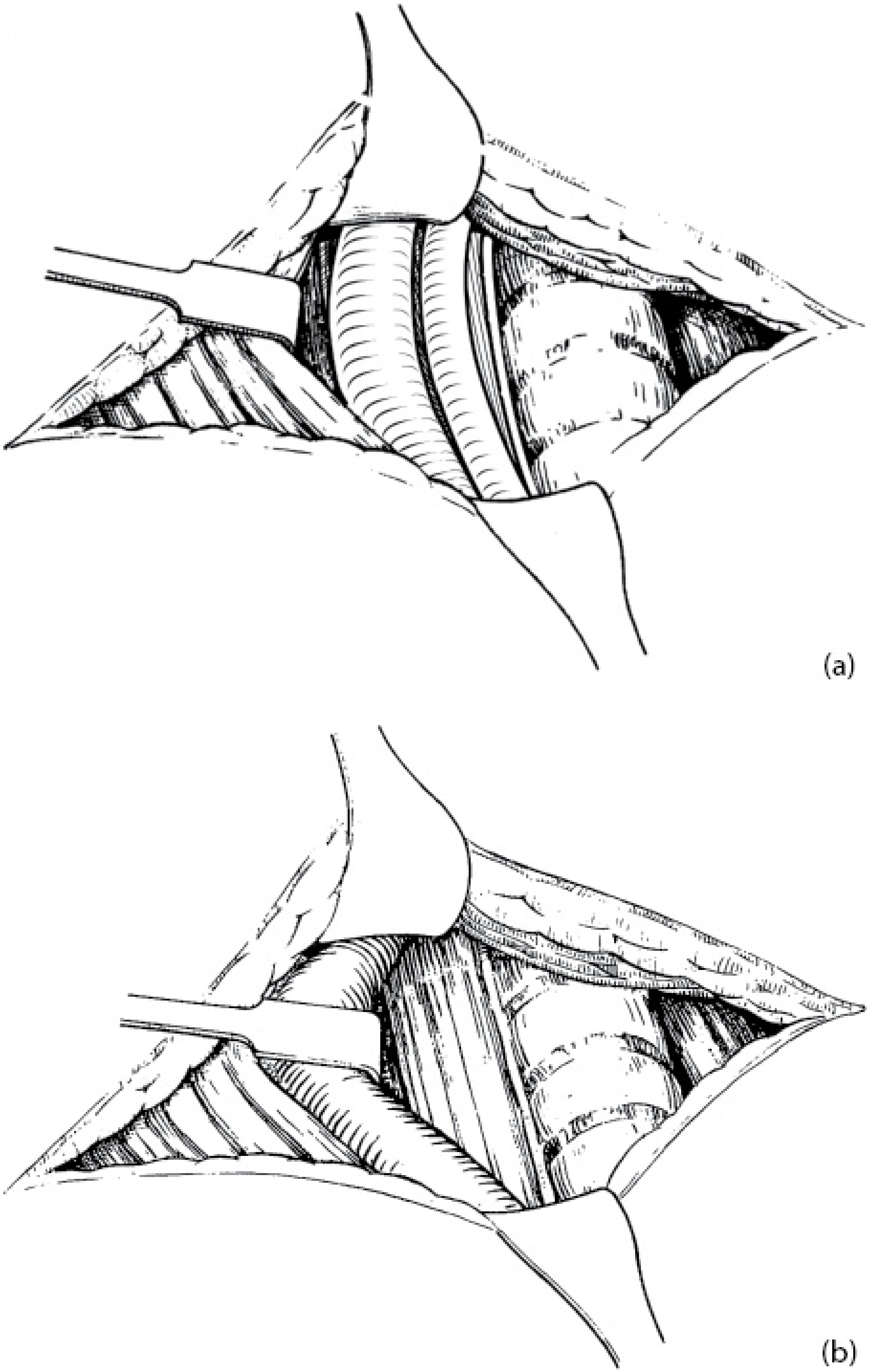




11.6

CREATION OF THE CERVICAL ESOPHAGOSTOMY

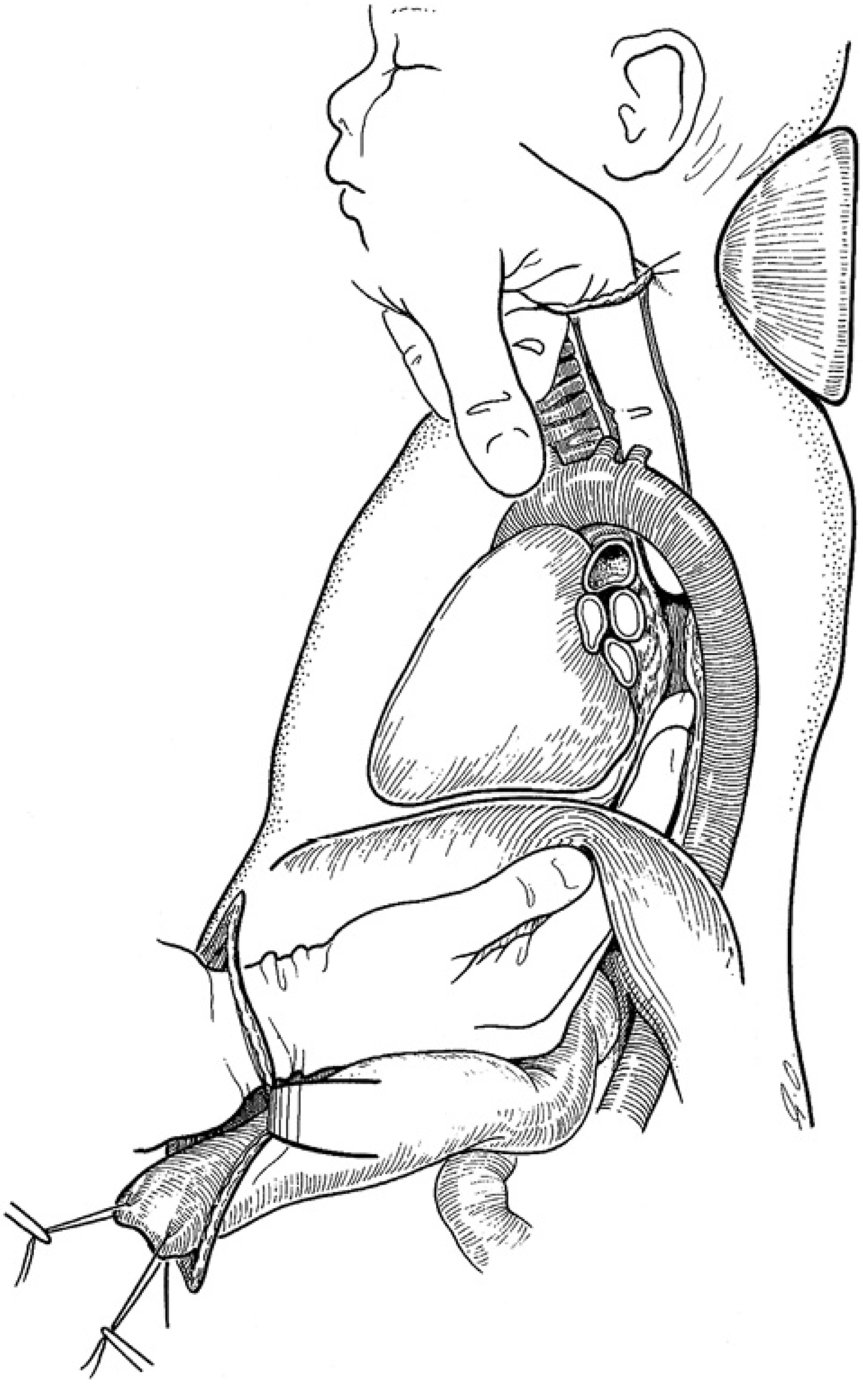
A right (or left) skin crease incision is used about 1 cm above and parallel to the clavicle. The incison is deepened through the platysma and the clavicular head of the sternocleidomastoid is divided. The sternohyoid may be retracted or divided to expose the carotid sheath (containing medially the common carotid artery and laterally the internal jugular vein and vagus nerve at the back) (**Figure 11.7a**). This is retracted laterally to develop the space between the centrally placed trachea and, behind it, the esophagus (**Figure 11.7b**). The recurrent laryngeal nerve lying in the groove needs explicit preservation. Mobilize and sling the esophagus, dissecting close to its muscular wall and separating it from the back of trachea. Continue the dissection into the cervical inlet freeing the blind upper pouch.



11.7a,b

PREPARING THE POSTERIOR MEDIASTINAL TUNNEL

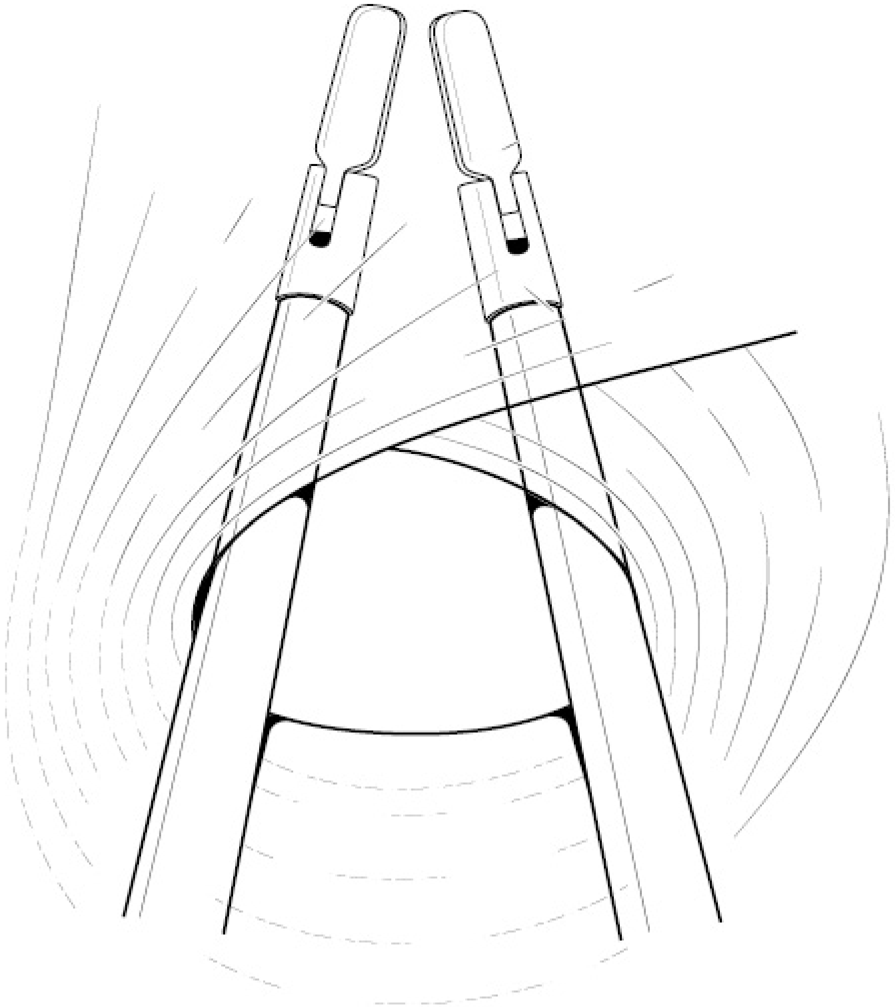
A plane of dissection between the membranous posterior surface of the trachea and the prevertebral fascia is established, and a tunnel is created into the superior mediastinum by blunt dissection immediately in the midline (**Figure 11.8**).



11.8

A similar tunnel is fashioned from below in the line of the normal esophageal route, by means of blunt dissection through the esophageal hiatus in the posterior mediastinal space posterior to the heart and anterior to the prevertebral fascia.

In the case of a laparoscopic approach, the posterior mediastinum is visualized from below using a zero-degree camera inserted via the Hasson cannula. Space is created in the posterior mediastinum using a “two-stick” triangulation maneuver with blunt dissection by two laparoscopic graspers advanced through the esophageal hiatus or by monopolar hook diathermy (**Figure 11.9**). Occasionally, if previous thoracic surgery has been performed, it may be necessary to mobilize the esophagus in the mediastinum via a thoracoscopic approach.



11.9

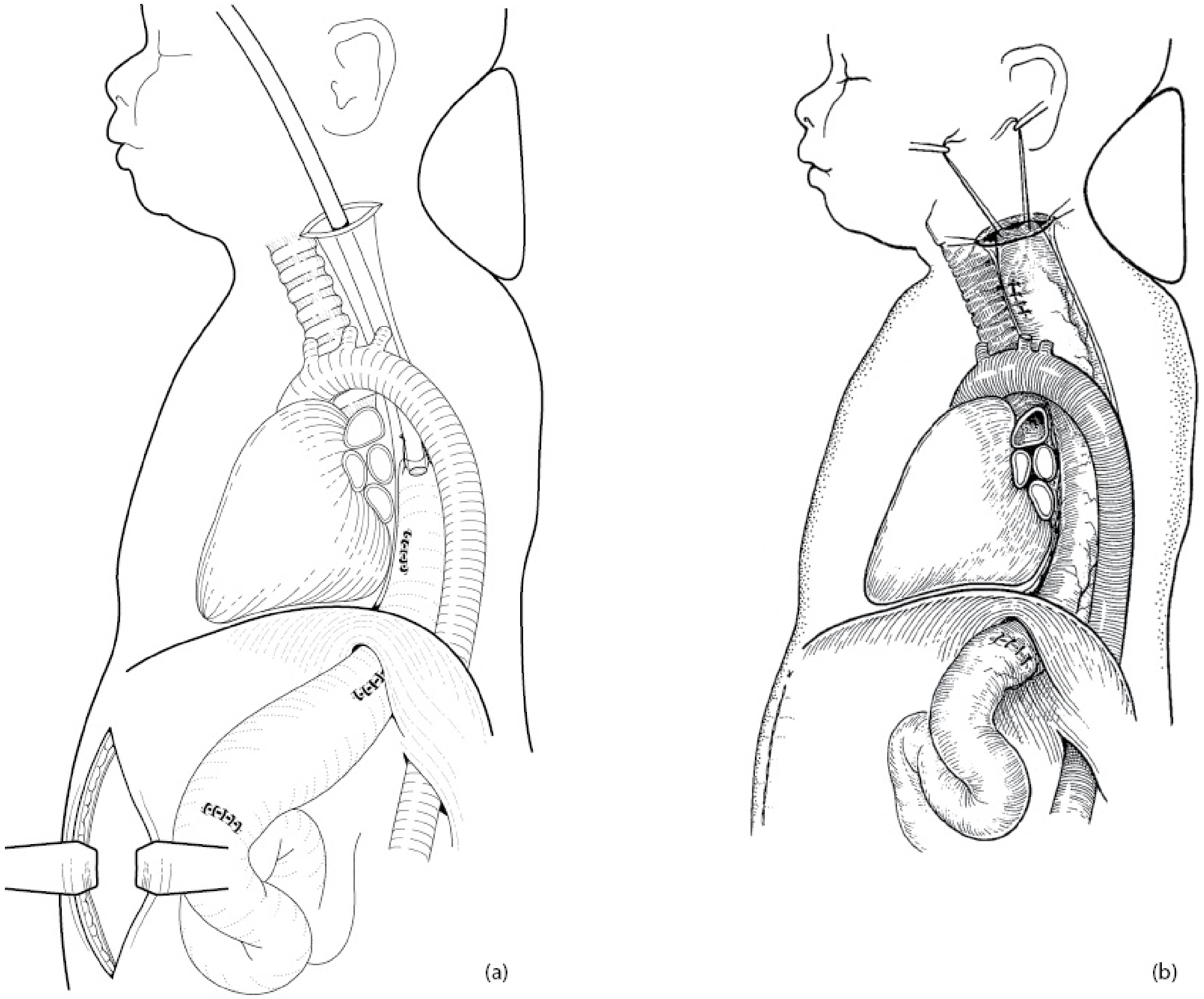
The mediastinal tunnel is further dilated using a long blunt-ended forceps and then increasing sizes of a Hegar dilator™ (Seward Thackray, Gwent, UK) to accommodate the transposed stomach. The laparoscopic camera is advanced above the hiatus and the Hegar dilators are introduced through the cervical approach and are seen emerging into the upper mediastinum.

When continuity of the superior and inferior posterior mediastinal tunnels has been established, the space to be occupied by the stomach is developed into a tunnel of two to three fingers' breadth.

There will be occasions when fashioning of the posterior mediastinal tunnel by blind dissection is impossible or hazardous due to inflammation, fibrosis from previous surgery, or adhesions following previous perforation or caustic ingestion. In these circumstances, it is necessary to perform a lateral thoracotomy or a thoracoscopy and for the dissection to be carried out under direction vision.

TRANSPOSING THE STOMACH

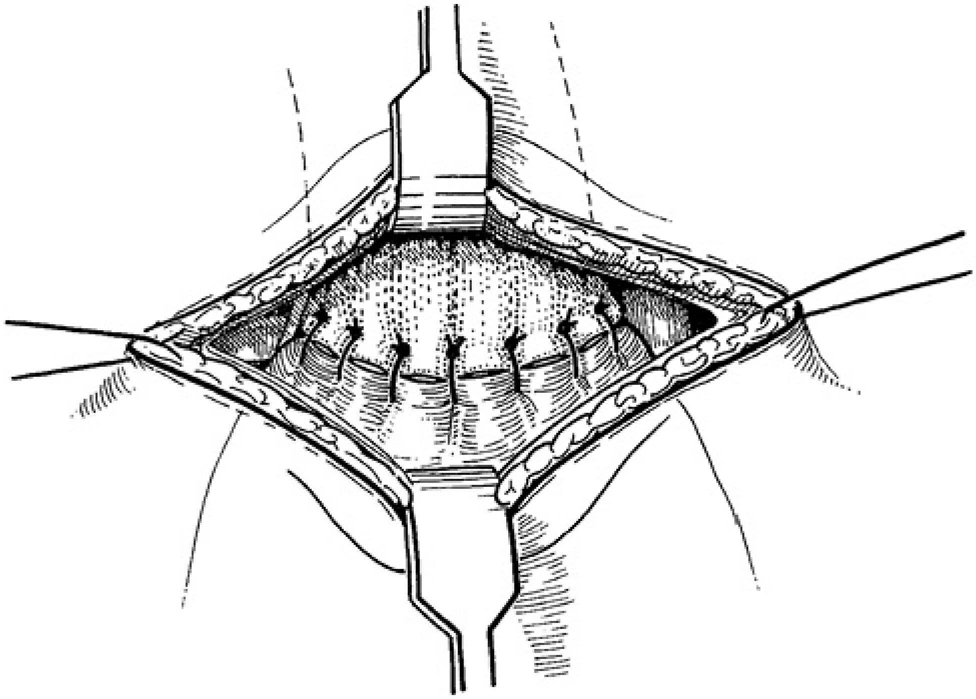
A large artery forceps is passed through the posterior mediastinal tunnel from the cervical incision to appear via the esophageal hiatus into the upper abdomen (**Figure 11.10a**). The two stay sutures on the fundus of the stomach are engaged to the forceps, which are then gently withdrawn, pulling the stomach up through the esophageal hiatus and the posterior mediastinal tunnel into the cervical incision (**Figure 11.10b**). Orientation of the fundus is checked by realigning the stay sutures in their correct position to avoid torsion of the stomach in the mediastinum.



11.10a,b

GASTROESOPHAGEAL ANASTOMOSIS

The transected end of the cervical esophagus is now anastomosed to the highest part of the stomach using a full-thickness single layer of interrupted 4/0 Vicryl™ sutures (**Figure 11.11**).

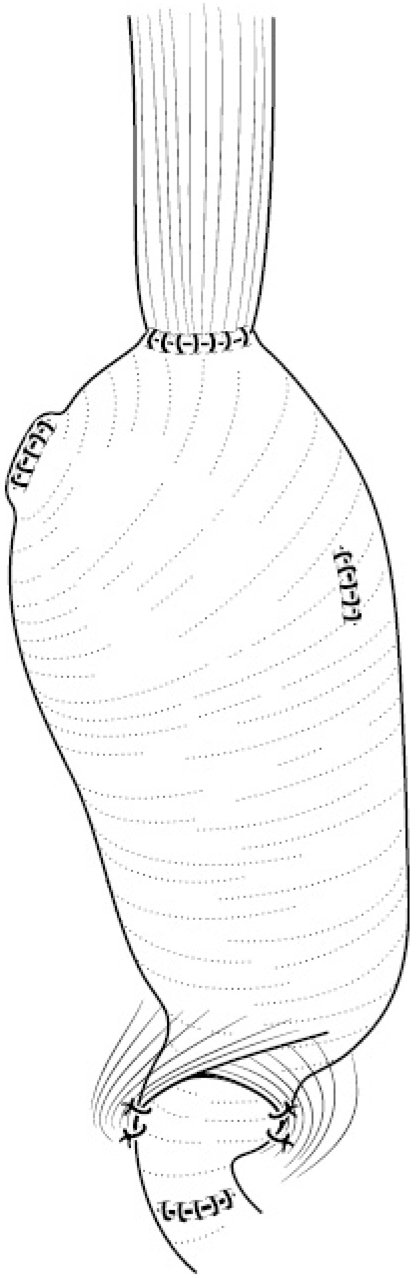


11.11

A 10–12 Fr transanastomotic nasogastric tube is inserted into the intrathoracic stomach before completing the anterior layer of the anastomosis. This is left on free drainage and aspirated at regular intervals to prevent acute gastric dilatation in the early postoperative period.

HIATAL DEFECT

It is important to prevent hiatal herniation by meticulous reapproximation of the diaphragmatic crura once the stomach has been pulled up and suturing of the antrum of the stomach to the diaphragmatic crura with 4/0 Vicryl□ or braided polyamide (Nurolon□) (**Figure 11.12**).



11.12

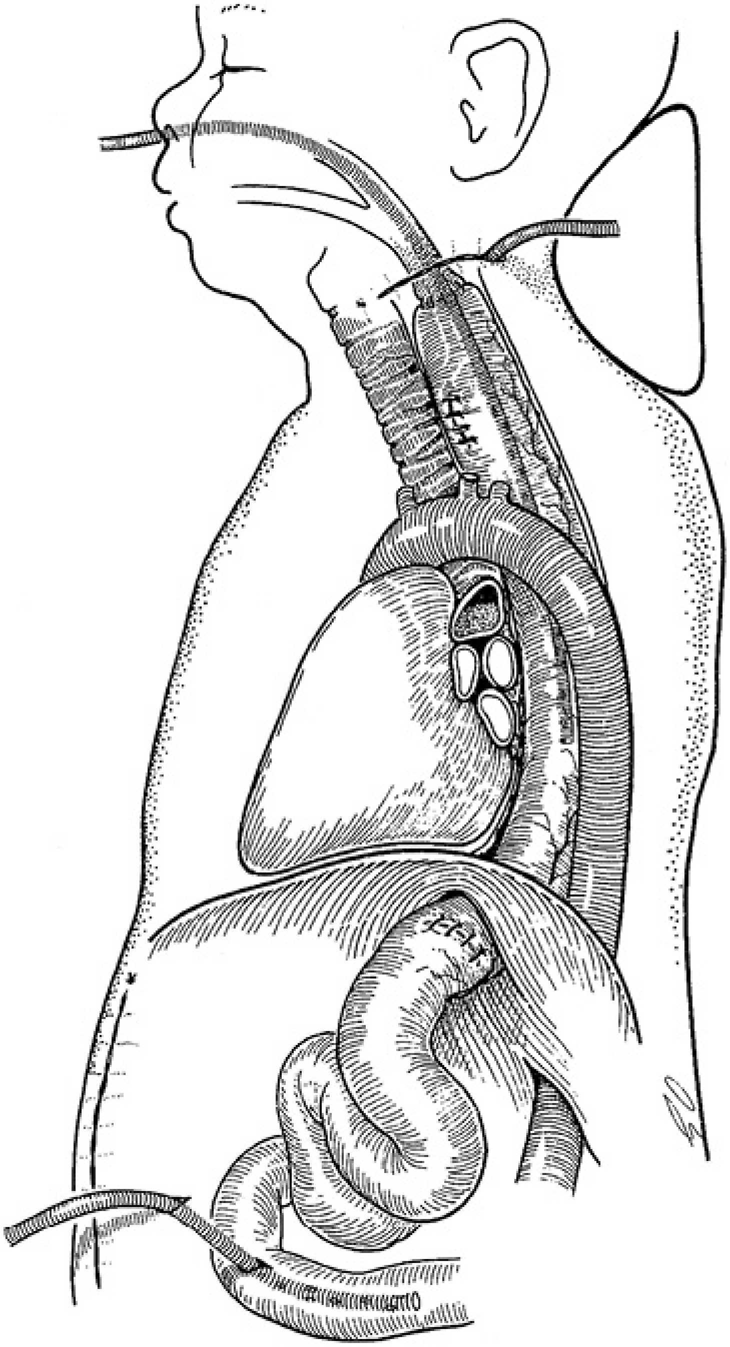
WOUND CLOSURE

The neck wound is closed in layers. The margins of the diaphragmatic hiatus are sutured to the antrum of the stomach with a few interrupted sutures, so that the pylorus lies just below the diaphragm. This also avoids hiatus hernias which have been described, particularly with the laparoscopic approach. If the chest is not opened, a chest drain is not required.

A fine-bore feeding jejunostomy has been found to be of considerable value in providing enteral nutrition in the first few weeks following gastric transposition, before full oral nutrition is established. This can be avoided in children with good sham feeding.

FINAL ANATOMY

The gastroesophageal anastomosis is shown in **Figure 11.13** in the cervical region with the nasogastric tube passing into the intrathoracic stomach. The pyloroplasty is below the diaphragm and a feeding jejunostomy tube is inserted for postoperative feeding. This is particularly important for infants with esophageal atresia who have previously not acquired the skill of oral feeding. The jejunostomy is a source of potential complications and it is probably wise to omit it in older children who have previously taken full oral nutrition (e.g. caustic strictures). A nasogastric tube is used, with the tip positioned in the middle of the mediastinum. If the chest is not opened, a chest drain is not required.



11.13

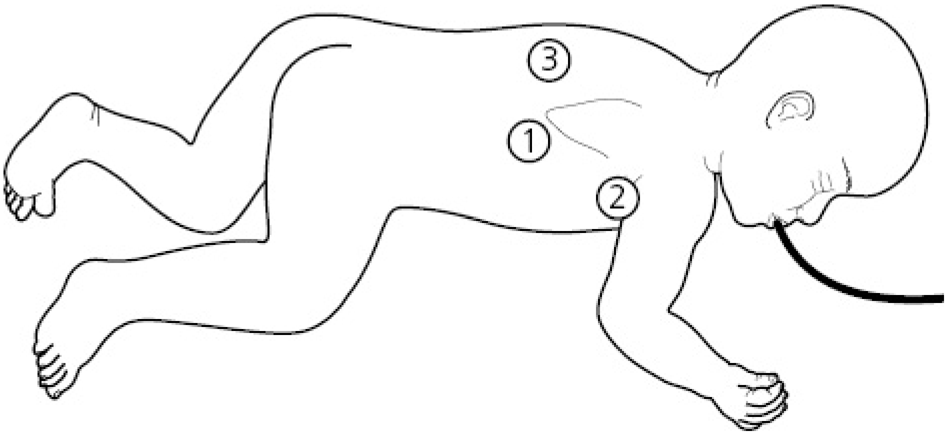
Alternative approach in cases of previous thoracic surgery or corrosive esophagitis

This approach may be used for patients who have undergone previous thoracic surgery or who have corrosive esophagitis (patient without cervical esophagostomy).

The laparoscopic procedure is performed first as described earlier. The position of the patient for laparoscopy is illustrated in **Figure 11.1**.

The thoracoscopy (or if necessary, thoracotomy) approach is performed.

The child is placed in a three-quarters left prone position (**Figure 11.14**). The surgeon is at the left side of the table with the screen opposite at the right upper end of the table. The assistant is at the surgeon's left side, and the scrub nurse at the bottom end of the table.



11.14

Three cannulas are inserted in the right chest in a V-shape. The first cannula (5 mm) is inserted just anterior to the angle of the scapula. A 30-degree telescope is inserted and the chest insufflated with CO_2 at 5 mmHg and flow of 2 L/min until the lung is collapsed. Following this, the CO_2 insufflation pressure can be reduced. A 5 mm port is inserted posterior to the scapula for the use of working instruments. A further 5 mm port is inserted in the axilla. The lung collapses or is retracted anteriorly to visualize the posterior mediastinum. Occasionally, if the lung does not collapse completely, it is necessary to introduce another 5 mm port posterior to the other two working ports to retract the lung anteriorly by using atraumatic forceps. Meticulous dissection is performed due to the adhesions in the mediastinum.

The minimally invasive procedure described above is challenging and should be performed by surgeons with experience in performing advanced laparoscopy and thoracoscopy.

POSTOPERATIVE CARE

Careful monitoring of vital functions is essential in the early postoperative period. There has been a fairly extensive dissection in the tissues posterior to the trachea, and edema may produce respiratory embarrassment. Elective nasotracheal intubation with assisted ventilation for a few days will simplify the postoperative course and reduce the incidence of respiratory proems.

Jejunal feeds are instituted on the second or third day after operation. The safest method of delivery of these feeds is by a slow continuous infusion rather than as a bolus, which can provoke a “dumping” effect. A contrast swallow is performed 5–7 days after surgery and, if no leak is identified at the anastomosis, oral feeding may be commenced. The cervical drain is removed when the integrity of the anastomosis has been demonstrated.

COMPLICATIONS

In the period 1981–2005, 192 gastric transpositions were performed at Great Ormond Street Hospital. Of these, 52% were via the posterior mediastinal route without thoracotomy. The mortality rate was 5.2%. Anastomotic leakage occurred at the esophagogastric connection in 12% and strictures developed in 19%. All leaks except one closed spontaneously and all strictures except three responded to endoscopic dilatation. Strictures were more common (38%) after caustic ingestion.

There are few reports on the laparoscopic-assisted gastric replacement of the esophagus in children. In 2007, Shalaby et al. published a series of 27 cases of post-corrosive esophageal stricture that were treated with laparoscopic-assisted transhiatal esophagectomy and gastric transposition. They reported an anastomotic leakage rate of 11% and a stricture rate of 15%. Recently, we noticed that the development of a large hiatus hernia appears to be more frequent in the laparoscopic approach compared to the more established open operation.

Delayed gastric emptying and dumping syndrome also occurred in some patients, but usually resolved spontaneously within a few months. Feeding problems and recurrent vomiting are commonly encountered in the early postoperative period, but they are generally transient.

OUTCOME

Good to excellent results are achieved in 90% of patients. Growth and development do not appear to be affected, and respiratory function is not impaired.

FURTHER READING

Hirschl RB, Yardeni D, Oldham K, et al. Gastric transposition for esophageal replacement in children: Experience with 41 consecutive cases with special emphasis on esophageal atresia. *Ann Surg* 2002; 236: 531–9.

Ng J, Loukogeorgakis SP, Pierro A, et al. Comparison of minimally invasive and open gastric transposition in children. *J Laparoendosc Adv Surg Tech A* 2014; 24: 742–9.

Orringer MB, Sloan H. Esophagectomy without thoracotomy. *J Thorac Cardiovasc Surg* 1978; 76: 643–54.

Shalaby R, Shams A, Soliman SM, et al. Laparoscopically assisted transhiatal esophagectomy with esophagogastroplasty for post-corrosive esophageal stricture treatment in children. *Pediatr Surg Int* 2007; 23: 545–9.

Spitz L, Kiely E, Pierro A. Gastric transposition in children – a 21-year experience. *J Pediatr Surg* 2004; 39: 276–81.

Stanwell J, Drake D, Pierro A, et al. Paediatric laparoscopic assisted gastric transposition. Early experience and outcomes. *J Laparoendosc Adv Surg Tech* 2010; 20: 177–81.

Ure BM, Jesch NK, Sümpelmann R, Nustede R. Laparoscopically assisted gastric pull-up for long gap esophageal atresia. *J Pediatr Surg* 2003; 38: 1661–2.

12

Esophageal replacement with jejunal interposition

DAVID C. VAN DER ZEE

HISTORY

In the past any esophageal atresia that could not be anastomosed would be called long-gap atresia. In a recent Position Paper on long-gap esophageal atresia, the definition was clearly determined as those types of esophageal atresia that had no air on plain abdominal X-ray.

Reconstruction of the esophagus in long-gap esophageal atresia (types A and B) has always been a challenge. (More recently the results of thoracoscopic traction technique demonstrated that most cases can nowadays be managed by delayed primary anastomosis or even directly after birth without the need for a gastrostomy.) For cases where primary anastomosis is not possible, many different techniques have been developed over the years, indicating that reconstruction is not an easy procedure. In 1946, Reinhoff performed an intrathoracic jejunal replacement of the esophagus. Jejunal interposition for long-gap esophageal atresia was first described by Akiyama et al. in 1971, and was later adopted by Bax et al. The technique is demanding, but the results are encouraging, even in the long term.

PRINCIPLES AND JUSTIFICATION

The jejunum is ideally suited for esophageal replacement, because it maintains good isoperistalsis and has a growth rate similar to that of the normal esophagus. In contrast to other techniques, there is little or no reflux and there are no pulmonary sequelae.

With good nursing care and a sump drain in place, the proximal esophagus can be drained adequately in the period before the reconstruction and a cervical esophagostomy should be avoided. A prior esophagostomy necessitates that a cervical proximal anastomosis will have to be performed when a reconstruction is undertaken. The crucial part of the procedure is meticulous dissection of the jejunal pedicle graft. Ideally, the proximal anastomosis is placed in the thorax.

The procedure commences with a right-sided thoracoscopy to obtain an accurate assessment of the proximal and distal esophagus and to determine whether a primary anastomosis is possible. If a primary anastomosis or thoracoscopic traction is not possible, a jejunal interposition is indicated, the thoracoscopic procedure is terminated, and the patient is repositioned into a supine position for a midline laparotomy and preparation of the jejunal pedicle graft. If the graft is judged to be of adequate length, a path is created retrocolic through the lesser omentum and esophageal hiatus. It is vitally important to ensure that the pedicle is not twisted when bringing it up into the thorax. After the abdominal wound has been closed again, a thoracotomy is performed, and the graft is tailored to its proper size. Both the proximal and distal anastomoses are preferably made intrathoracically.

PREOPERATIVE

Assessment and preparation

The first step in a neonate with a long-gap esophageal atresia is usually the placement of a gastrostomy for feeding. During this procedure, a tracheobronchoscopy is carried out to exclude a proximal fistula. A proximal fistula usually prevents the proximal esophagus from increasing in length. Through the gastrostomy, the length of the distal esophagus can be determined, either with bougies or by contrast study. An intestinal contrast study to determine the length of the small intestine is performed just prior to the interposition procedure.

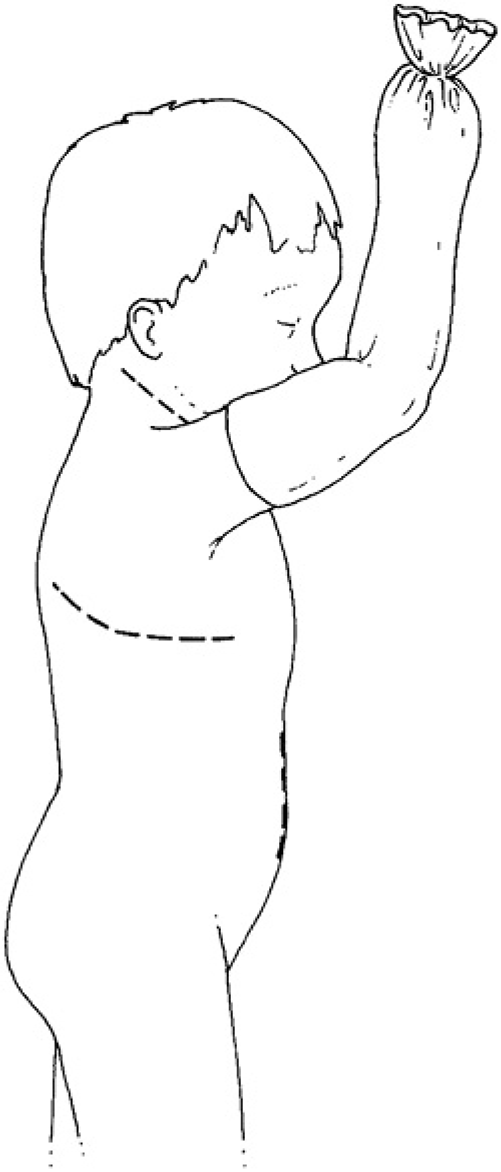
No specific preoperative bowel management measures are necessary.

Anesthesia

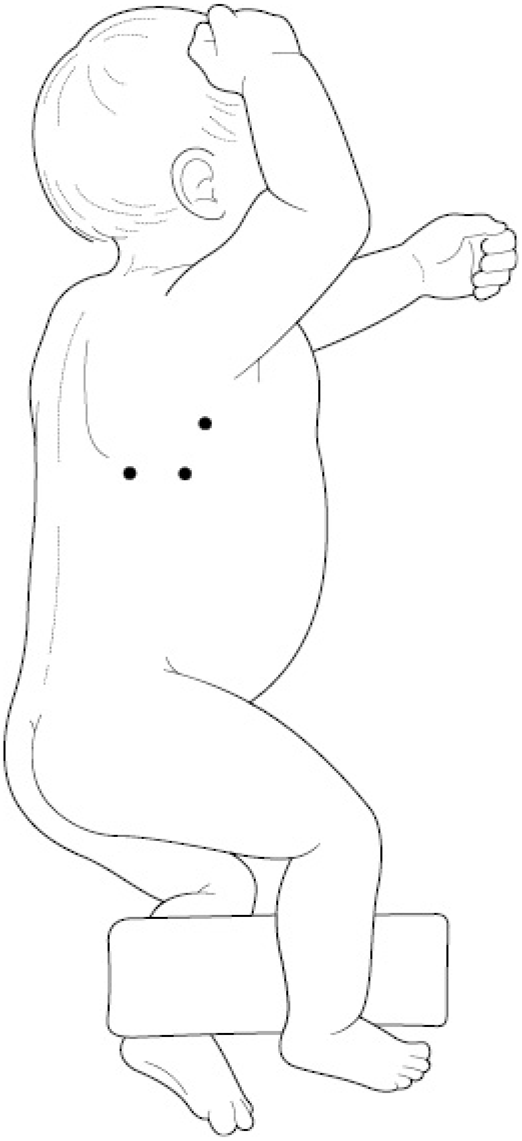
Jejunal interposition is accomplished under general anesthesia. An epidural catheter for intra- and postoperative pain management is inserted. An arterial line is placed for sampling during the procedure and postoperatively. Because of the length of the procedure, a urine catheter is introduced.

OPERATION

The patient is positioned in either a left lateral decubitus position for thoracotomy (**Figure 12.1**) or a left three-quarter left prone position for thoracoscopy (**Figure 12.2**). Exploration and mobilization of the two ends is performed (**Figure 12.3**). If it is determined that a primary or delayed primary is not possible, the distance between the two ends is measured, in order to prepare the jejunal pedicle graft. The trocars are taken out, the patient is positioned in a supine position and a midline laparotomy is undertaken. When in place, the gastrostomy is taken down. The proximal jejunal loops are inspected for anomalies in their anatomy that might preclude the dissection.



12.1



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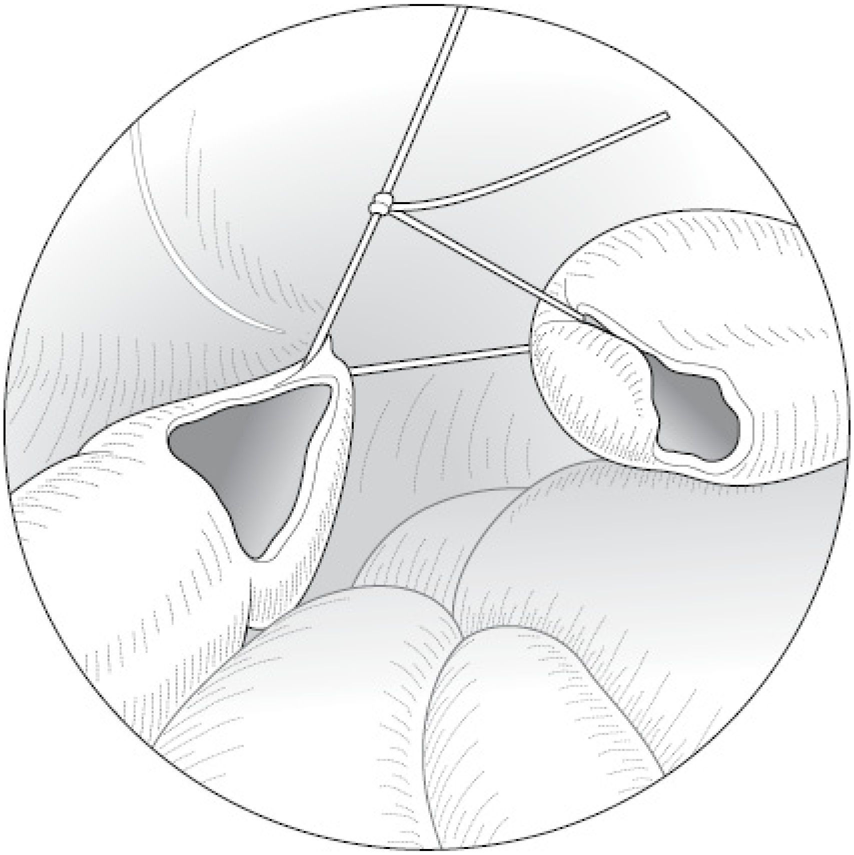


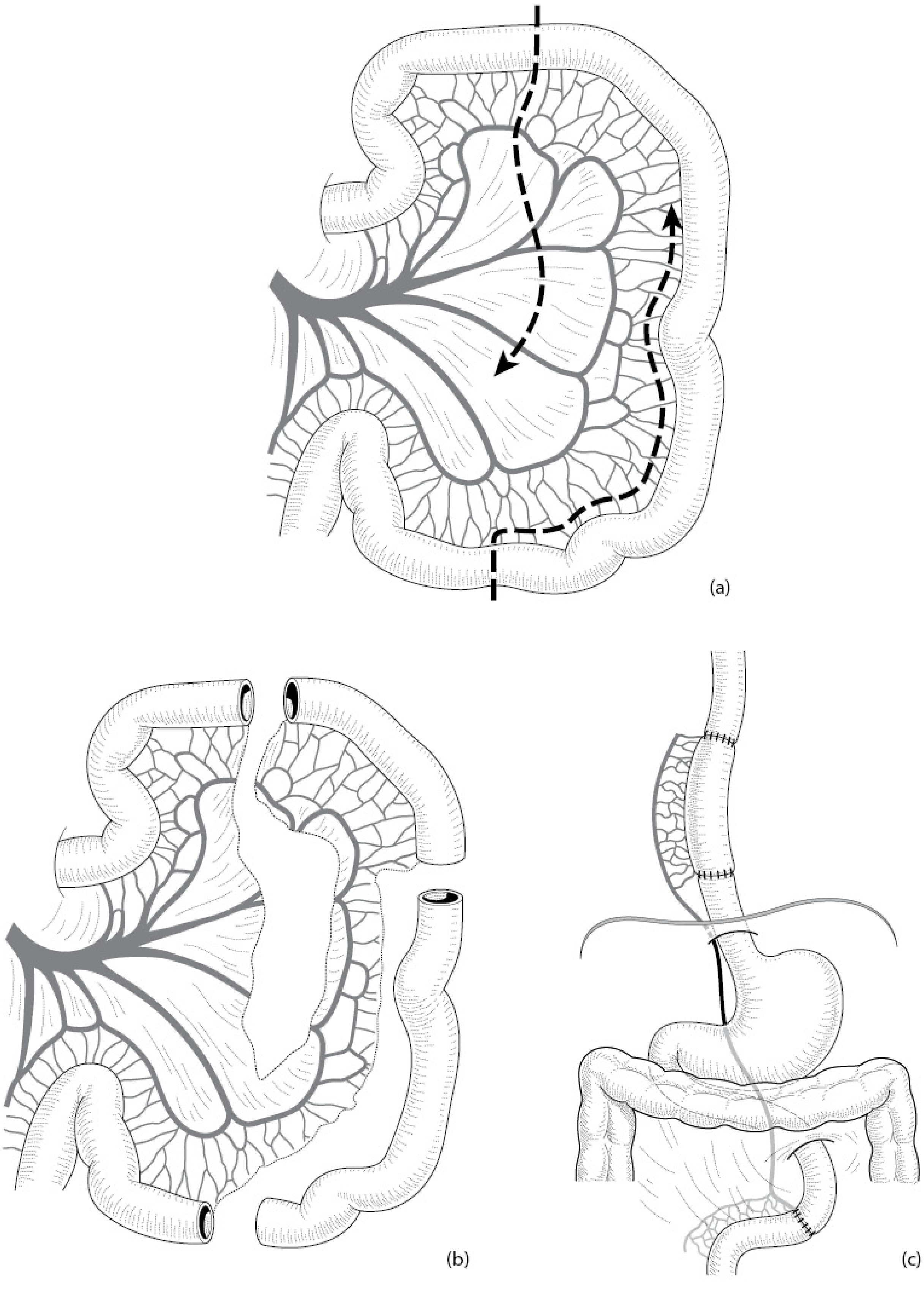
12.3

The next step is to prepare everything for transposition of the graft into the thorax. This entails mobilization of the esophageal hiatus to allow the passage of the graft into the thorax. Usually the short gastric vessels are taken down in order to facilitate the entrance to the lesser omentum and hiatus. The hiatus is dilated using Hegar dilators up to H14.

The proximal jejunum is transected approximately 3–5 cm from the ligament of Treitz and two mesenteric branches are severed centrally to gain length of the pedicle. The length of the pedicle is measured to determine if sufficient length has been obtained before the distal jejunal end is transected just proximal to the third mesenteric branch. The two ends of proximal and distal jejunum are anastomosed.

Only the most proximal 3–5 cm of jejunum will be used for the interposition and the rest of the mobilized jejunal is carefully dissected from its vasculature at the level of the jejunal serosa and resected (**Figure**

12.4a). Initially, the distal portion of jejunum may appear somewhat bluish due to the ‘steal’ of circulation in the proximal part of the jejunum, but as dissection progresses the circulation improves and the final 3–5 cm of jejunum remaining on the pedicle resumes good vascularity (**Figure 12.4b**). It is important to keep an eye on the position of the vasculature at all times to avoid twisting or strangulation of the vascular stalk.


(a)
(b)
(c)

12.4a–c

An opening is now made in the mesentery of the transverse colon somewhat on the left side to pass the jejunal graft with its pedicle through in a comfortable way. The graft and its blood supply are closely observed at all times while it is passed through the lesser omentum and through the esophageal hiatus. The jejunal graft is carefully placed in the lower thorax.

Because time now plays an important role, the abdomen is closed provisionally.

The patient is repositioned in a left lateral decubitus position and a thoracotomy is performed. After retraction of the lung, the pedicled graft is located and carefully stretched into the thorax, carefully maintaining the vasculature (**Figure 12.4c**). The definitive length of the graft is determined and adjusted if necessary. First the proximal end of the jejunal graft is anastomosed to the proximal esophagus using 5/0 Vicryl® and, after that anastomosis has been completed, the distal portion can be adjusted for anastomosis to the distal esophagus. As the distal esophagus is hypoplastic in many instances, the distal esophagus is opened obliquely to obtain an adequate diameter for the anastomosis. The anastomosis is made using 5/0 Vicryl® interrupted sutures.

A transanastomotic tube is passed into the stomach. A thoracic drain is left *in situ* for the first few postoperative days.

After closure of the thorax, the patient is turned back into the supine position for refashioning of a gastrostomy and final closure of the laparotomy wound.

POSTOPERATIVE CARE

A contrast swallow is performed on postoperative day 5. If there is no leakage, oral feeding can be initiated.

If the child returns with feeding difficulties, a contrast study can exclude anastomotic strictures. Sometimes when the distal esophagus is only a small bud there may be dysphagia, requiring dilatation, but usually swallowing problems resolve in due course.

OUTCOME

Although the procedure is demanding, there has been no graft loss in our series. Twenty-seven children received a jejunum interposition between 1988 and 2009, of whom 22 had long-gap esophageal atresia (eight had a proximal fistula), three had caustic burns, and two had severe peptic strictures. Five patients developed an anastomotic leak (four thoracic and one abdominal). On follow-up, four children had complaints of reflux for which they were treated with antireflux medication. Five children occasionally experience functional stenosis at the distal anastomosis that responds well to domperidon. Two children are treated for upper airway complaints.

More recently, a comparative study on gastric pull-up and jejunal interposition demonstrated a more favorable outcome for jejunal interposition. Also, fewer respiratory problems were encountered.

The major advantage of jejunal pedicle grafts is that they grow at the same speed as the native esophagus, redundancy does not occur, and the grafts display peristalsis, thus facilitating good passage of solid food.

FURTHER READING

Akiyama H, Ishii K, Kodaira Y, et al. Esophageal reconstruction with a pedicled jejunum in esophageal atresia (type A). *Shujutsu* 1971; 25: 711–19.

Bax KN. Jejunum for bridging long-gap esophageal atresia. *Semin Pediatr Surg* 2009; 18: 34–9.

Bax KN, Roskott AM, van der Zee DC. Esophageal atresia without distal tracheoesophageal fistula: High incidence of proximal fistula. *J Pediatr Surg* 2008; 43: 522–5.

Bax NM, van der Zee DC. Jejunal pedicle grafts for reconstruction of the esophagus in children. *J Pediatr Surg* 2007; 42: 363–9.

Gallo G, Vrijlandt EJ, Arets HG, et al. Respiratory function after esophageal replacement in children. *J Pediatr Surg* 2017; 52(11): 1736–41.

Gallo G, Zwaveling S, van der Zee DC, et al. A two-center comparative study of gastric pull-up and jejunal interposition for long gap esophageal atresia. *J Pediatr Surg* 2015; 50(4): 535–9.

van der Zee DC, Bagolan P, Faure C, et al. Position Paper of INoEA Working Group on Long-Gap Esophageal Atresia: For better care. *Front Pediatr* 2017; 5: 63.

van der Zee DC, Gallo G, Tytgat SH. Thoracoscopic traction technique in long gap esophageal atresia: Entering a new era. *Surg Endosc* 2015; 29(11): 3324–30.

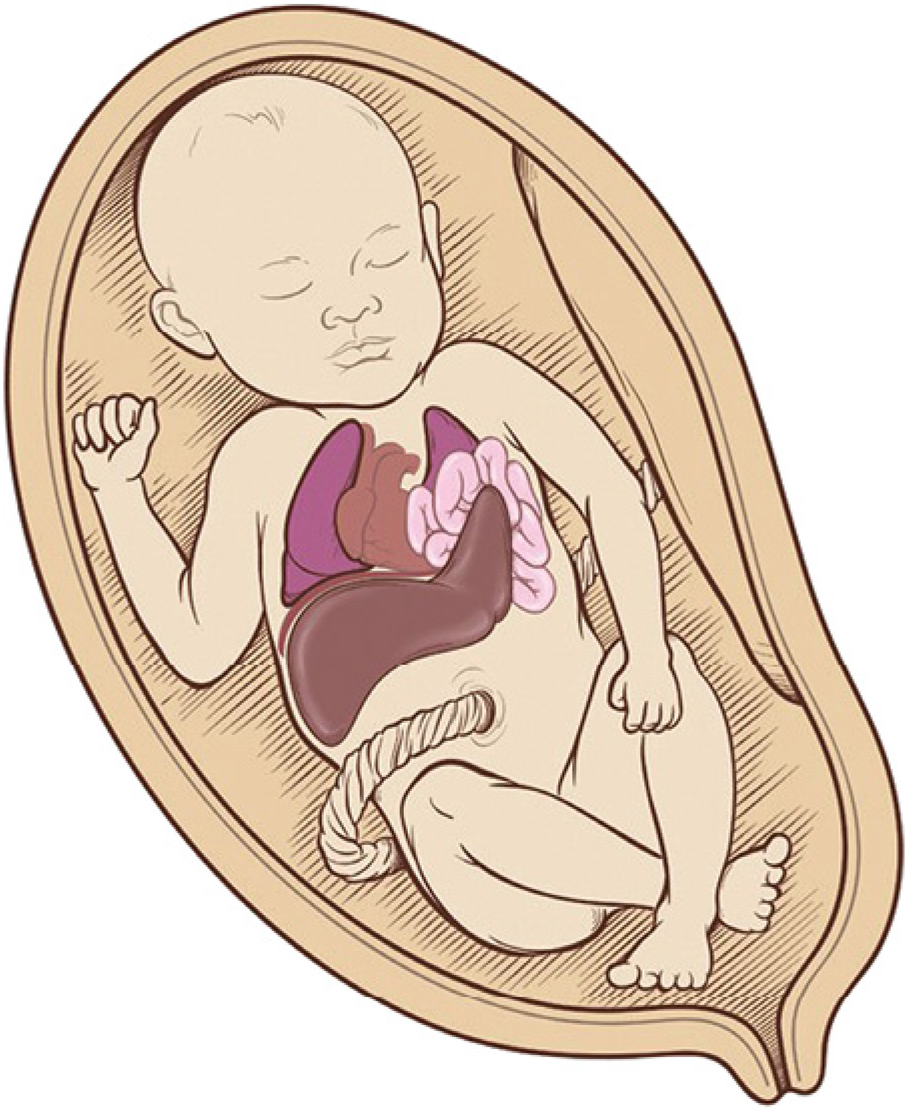
13

Congenital diaphragmatic hernia

ERIN E. PERRONE AND GEORGE B. MYCHALISKA

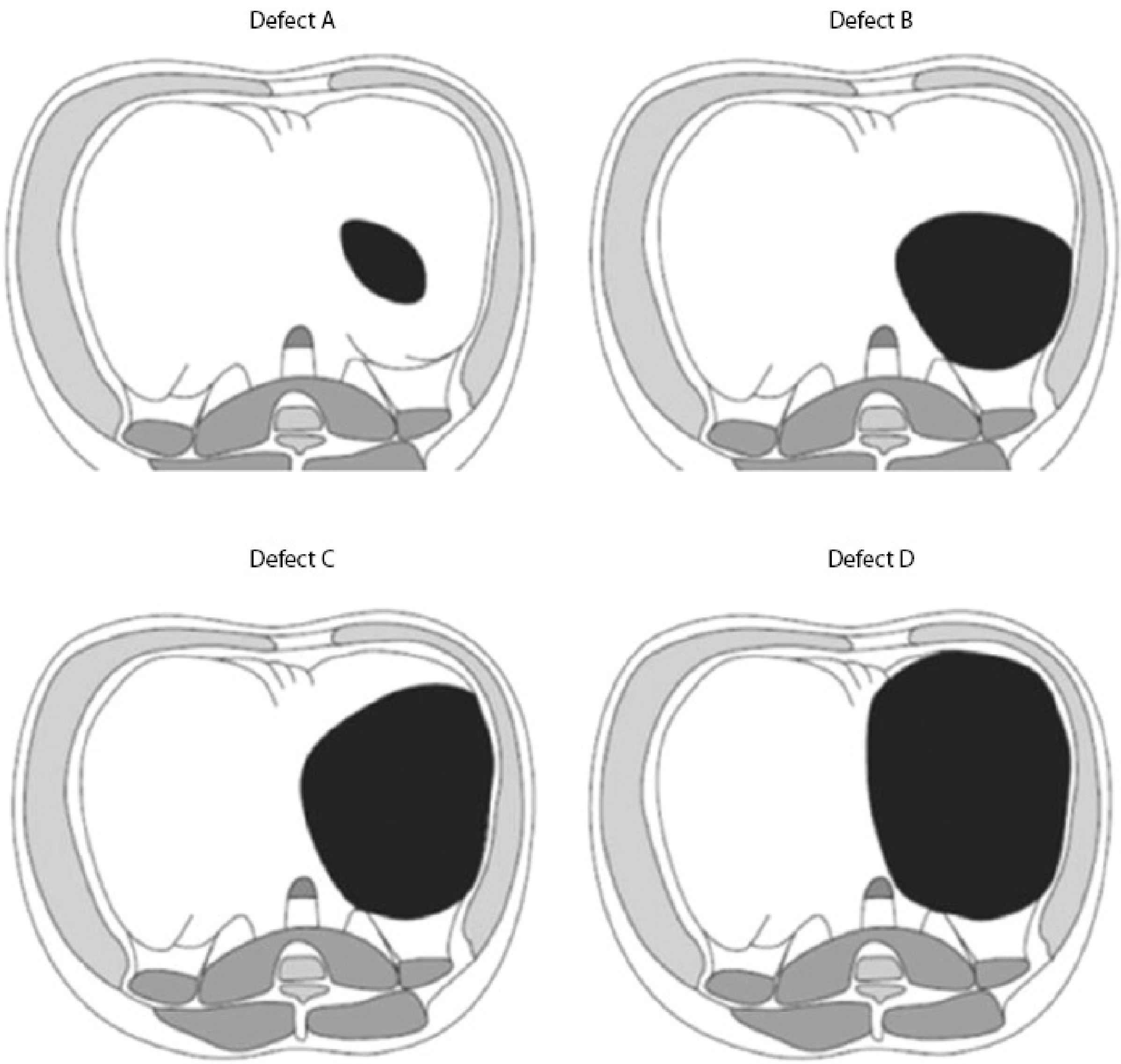
BACKGROUND

Congenital diaphragmatic hernia (CDH) occurs in up to 1 in 2500 live births. It is a field defect, involving the lungs, foregut, and chest wall, resulting from failure of the diaphragm to separate the pleuroperitoneal canal into the thorax and abdomen before the midgut returns through the umbilicus. Consequently, the abdominal viscera translocate into the chest in the first trimester when the lungs are at a vulnerable lung bud or glandular stage (**Figure 13.1**). The defect is typically located posteriolaterally, named Bochdalek hernia, and most of the chapter will focus on this type. Information specifically on Morgagni hernias which are anteriomedial defects is at the end of the chapter.



13.1 Illustration of infant with CDH. (Image owned by Michigan Medicine and printed with permission.)

Both lungs are involved, the ipsilateral greater than the contralateral lung. Affected infants are born with a complex combination of pulmonary hypoplasia and pulmonary hypertension. There is decreased surface area available for gas exchange, compromised bronchiolar and pulmonary vascular beds, and muscular hypertrophy of the intra-acinar arterioles. Despite significant advances in neonatal intensive care and pediatric surgical care, a cohort of affected infants does not survive. The most recent data from the Congenital Diaphragmatic Hernia Study Group (CDHSG) reports an overall survival of 71%, with survival lower in the prenatally diagnosed group (65%) when compared to the postnatally diagnosed group (83%). The CDHSG has developed a classification system based on defect size, with A being the smallest and D the largest (**Figure 13.2**). The system correlates with degree of pulmonary hypoplasia and severity. Mortality rate increases with defect size and is reported as 0.6%, 5.3%, 22.6%, and 45.6% in groups A, B, C, and D respectively. Pulmonary, neurologic, and gastrointestinal morbidity is also increased with defect sizes C and D.

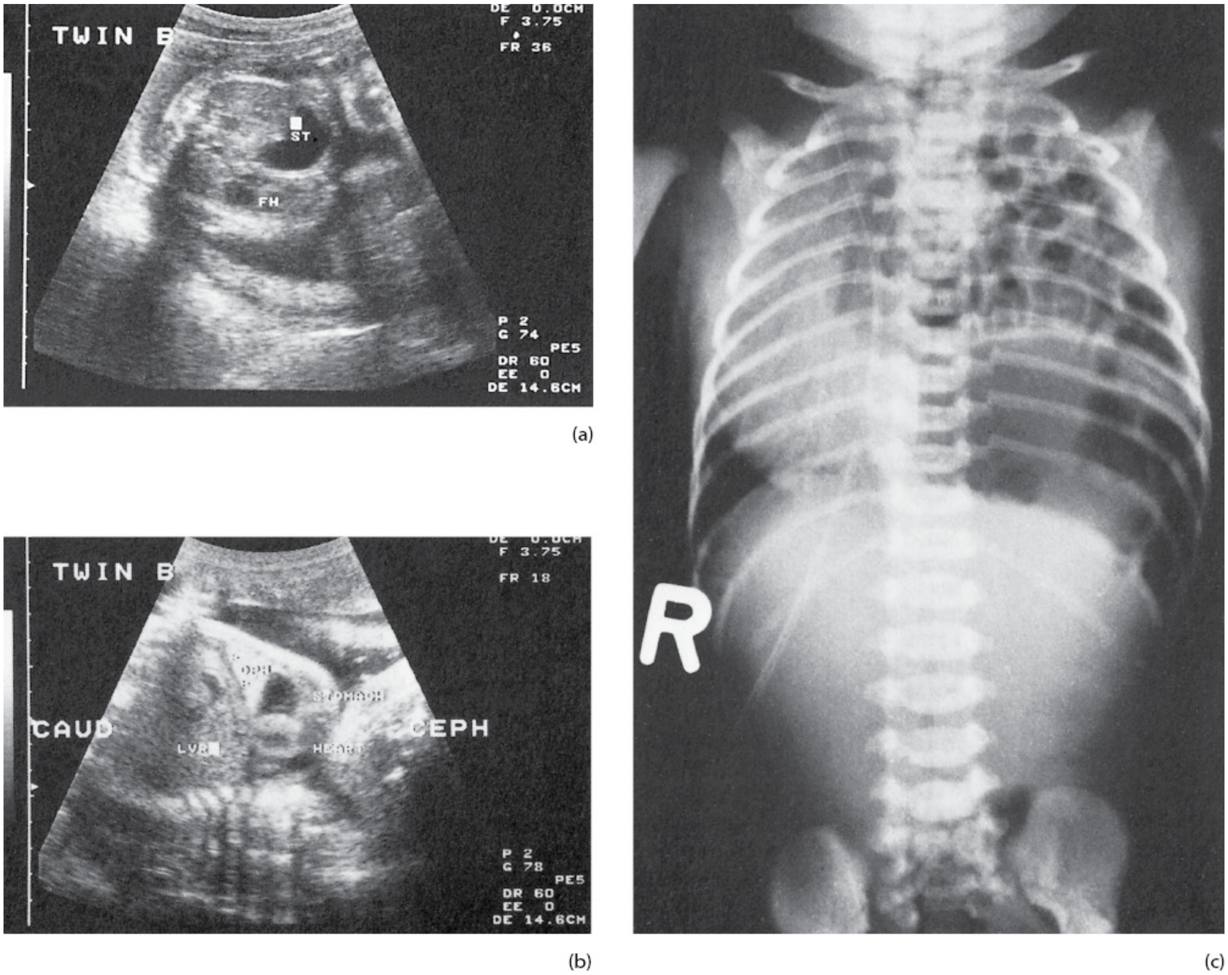


13.2 CDHSG Staging System. Defect A = surrounded by muscle; Defect B = small <50% of chest wall devoid of diaphragm tissue; Defect C = >50% of chest wall devoid of diaphragm tissue; Defect D = complete or near complete absence of diaphragm. There is no defect diagram for right-sided defects so the CDHSG recommends applying the above descriptions and reversing the diagrams (i.e. using mirror images) to determine the size of a right-sided defect.

DIAGNOSIS

The majority of infants with CDH are diagnosed antenatally (68%). Ultrasound may show bowel and/or liver in the chest, the heart and stomach in the same plane, or mediastinal shift (**Figure 13.3a–c**).

Ultrasound can give estimates of disease severity by calculating observed-to-expected lung to head ratio (o/e LHR) with the most severe disease having a calculated o/e LHR <25%. Fetal magnetic resonance imaging (MRI) has also become a useful adjunct to differentiate CDH from lung bud anomalies. Fetal MRI can estimate observed-to-expected total fetal lung volumes (o/e TFLVs), prognosticate survival for prenatal counseling, and evaluate the degree of liver herniation within the chest.


TWIN B
F 3.75
FR 36
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EE 0
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(a)
TWIN B
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STOMACH
CAUD
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LVR
HEART
P 2
G 78
PES
DR 60
EE 0
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(b)
R
(c)

13.3a–c

CDH infants are typically near-term and present immediately with respiratory distress, tachypnea, grunting, and cyanosis. Physical examination reveals a scaphoid abdomen, asymmetric respiration, and a barrel chest. Chest radiography confirms the diagnosis, showing multiple gas-filled bowel loops above the diaphragm, contralateral mediastinal shift, and a paucity of abdominal bowel gas.

PREOPERATIVE

Resuscitation

With antenatal diagnosis, the mother and fetus require transfer to an experienced center capable of appropriate neonatal, surgical, and extracorporeal membrane oxygenation (ECMO) care. If diagnosed postnatally, the infant is stabilized and transferred to the same type of specialized center for management and operative repair. Surgical intervention is not an emergency. Multiple studies demonstrate the benefit of medical stabilization over several days followed by elective repair. Infants with pulmonary hypertension and pulmonary hypoplasia refractory to conventional therapy should be considered candidates for ECMO. Timing of repair for patients who require ECMO is controversial, with some advocating for early repair on ECMO and others advocating for delayed repair after weaning off ECMO.

Initial treatment starts with prompt intubation, nasogastric decompression, and vascular access. Bag-mask ventilation should be avoided to limit distension of the stomach and intestines in the chest. Umbilical artery catheterization allows accurate blood pressure monitoring, and blood sampling. Because of variable amounts of right-to-left shunting at the ductus arteriosis, blood gas monitoring from the umbilical artery is misleading. A preductal arterial catheter or pulse oximetry is essential in guiding therapeutic interventions and evaluating gas exchange potential.

Respiratory care must avoid muscle paralysis and allow spontaneous respiration to minimize barotrauma. Ventilator settings range from low rates and modest peak airway pressures to high frequency oscillatory ventilation. The prevention of secondary lung injury cannot be overemphasized and gentle ventilation strategies are emphasized with ideal peak inspiratory pressures <25 mmHg.

Pharmacologic support consists of ionotropes, to treat hypotension. Milrinone, dopamine, and dobutamine are commonly used in this setting. Despite its widespread use pre-ECMO, there is no data to support routine use of inhaled nitric oxide (iNO). Other medications aimed at treating pulmonary hypertension are sildenafil and prostacyclin, which have a role in select patients. ECMO should be considered with suprasystemic pulmonary hypertension and a preductal saturation of less than 85% using a gentle ventilation protocol.

Anesthesia

The airway should be controlled with orotracheal intubation. Single lung ventilation is unnecessary. Intravenous anesthesia is administered as needed and complemented by muscle paralysis and narcotics. Mechanical ventilation may be controlled throughout surgery with a pressure-cycled infant ventilator rather than the conventional anesthesia machine. Continuous oxygen saturation monitoring, both preductal and postductal, is critical. An arterial line should already be placed and accessible for intraoperative blood gases. A heating mattress is used, and the head and extremities are wrapped to minimize heat loss. Prophylactic antibiotics should be given preoperatively.

OPERATIONS

Traditional open repair is accomplished transabdominally. Thoracoscopic techniques are advocated for stable patients.

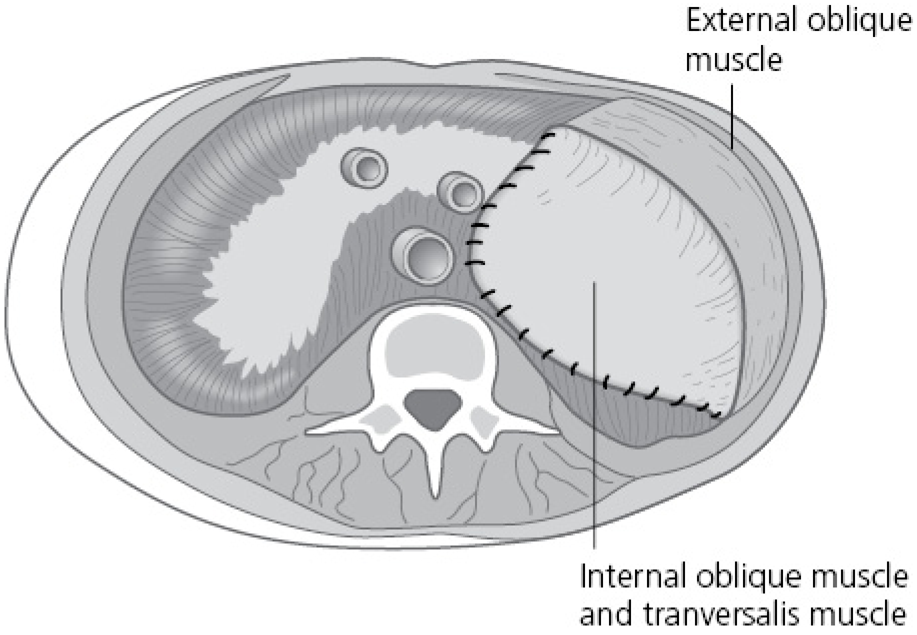
Transabdominal open repair

POSITION OF PATIENT

The infant is positioned supine, elevating the thoracolumbar spine. Both the upper abdomen and chest are prepared as the operating fields.

INCISION

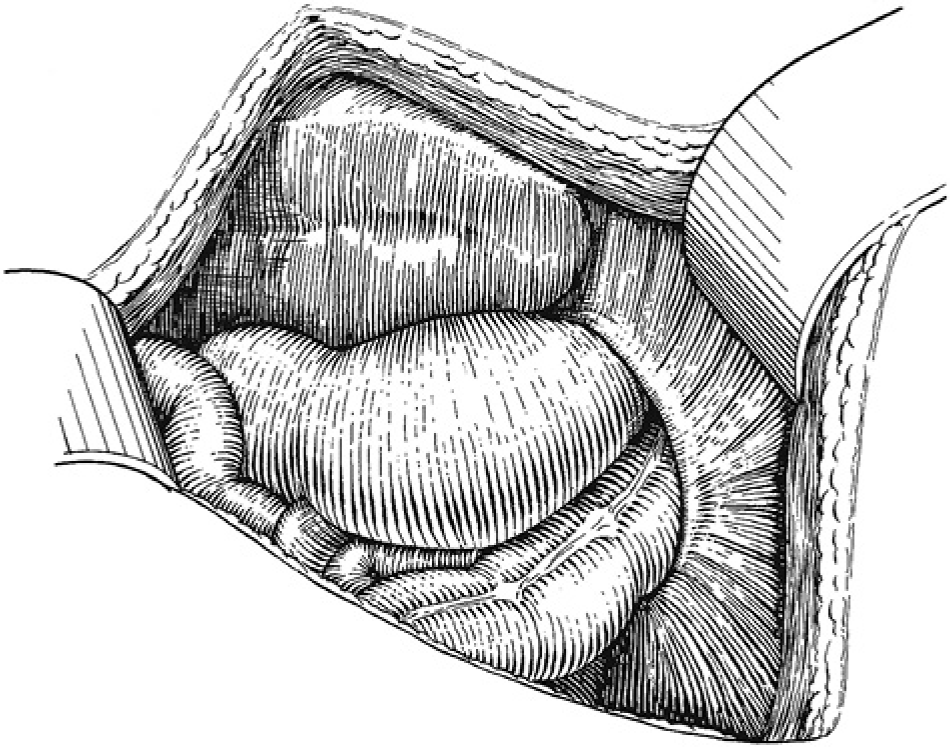
An ipsilateral subcostal incision is made 1 cm below the costal margin. Some surgeons advocate for a transversalis abdominis muscle flap instead of polytetrafluoroethylene (PTFE; Gore-Tex®) for larger defects (**Figure 13.4**). In these cases, it is important to remember to make the initial incision 2 cm below the costal margin in order to create the muscle flap.



13.4

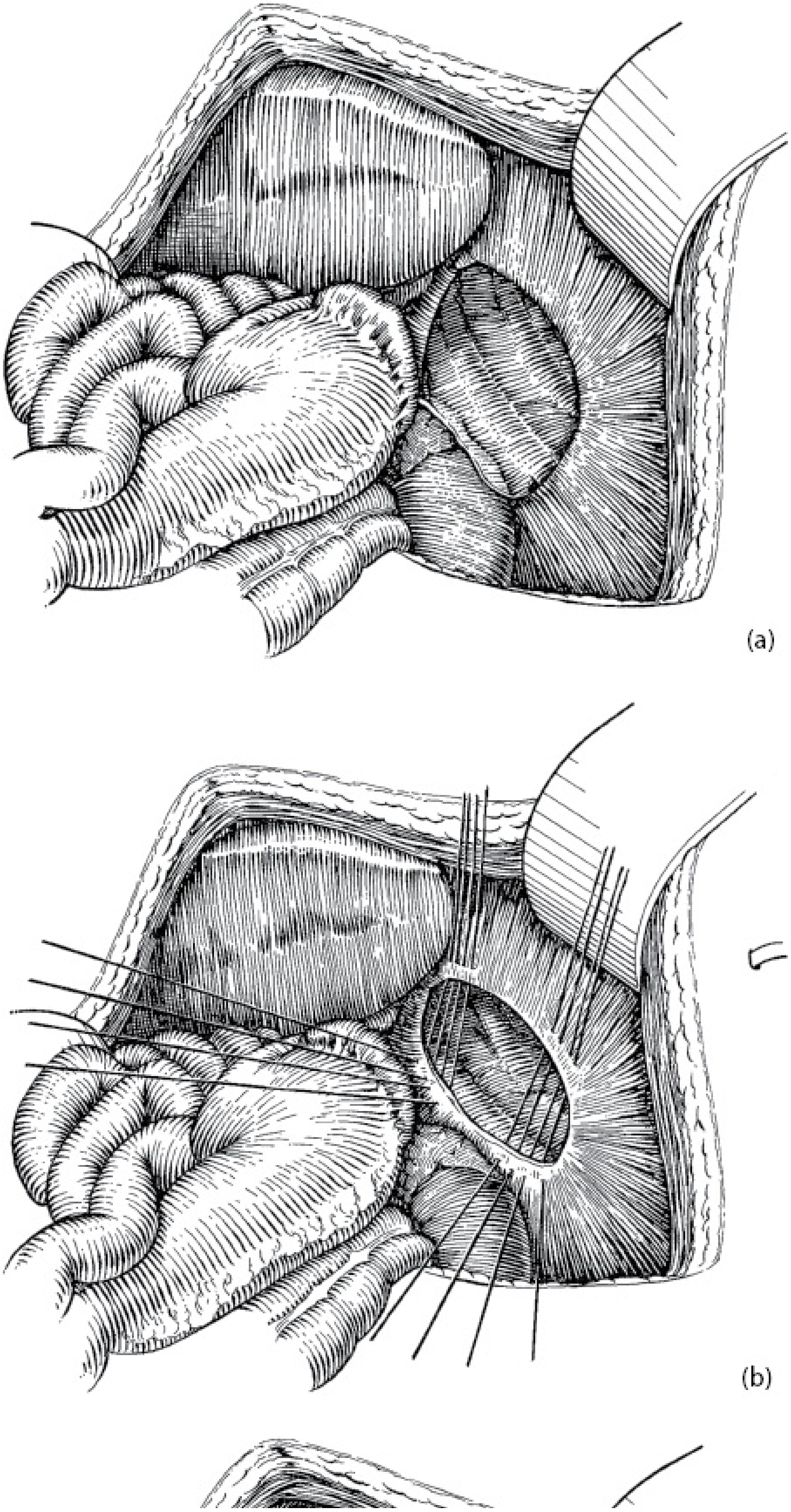
STEPS

The defect is exposed after the stomach is decompressed, and the viscera are reduced from the chest (**Figure 13.5**). The unfixed spleen and its attachments to the colon and pancreas are especially vulnerable. Reduction of the liver and spleen may be challenging, but can be accomplished by careful manual reduction using downward pressure from above.



13.5

If a true hernia sac exists, it should be excised to ensure proper healing of the defect (**Figure 13.6a**). The posterior rim can be identified by tracing the anterior rim medially. Its mesothelial covering is sharply incised and carefully mobilized (**Figure 13.6b**). Primary repair is accomplished with interrupted, simple, non-absorbable sutures with or without pledgets (**Figure 13.6c**).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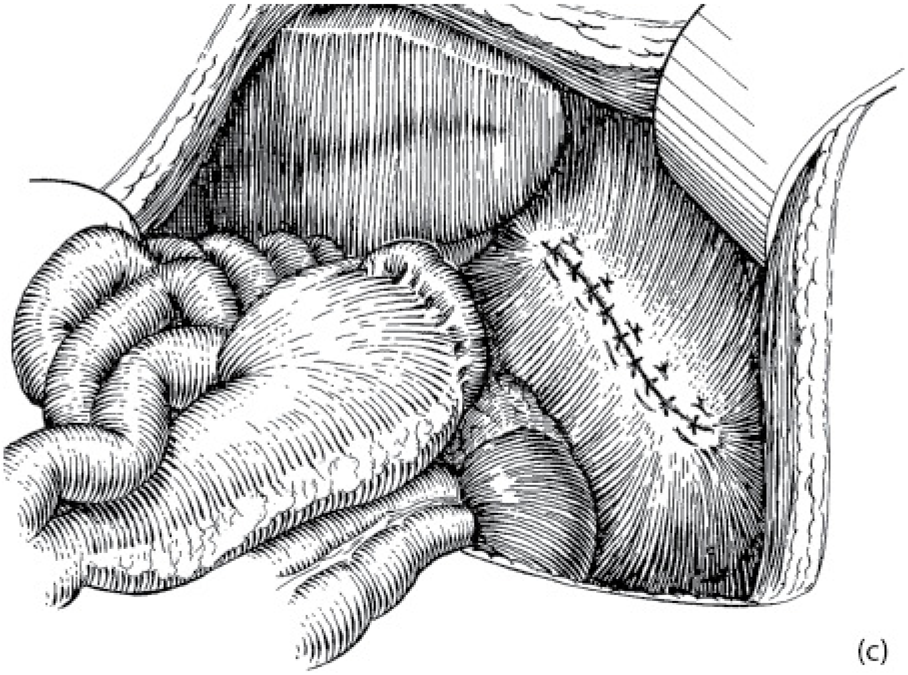
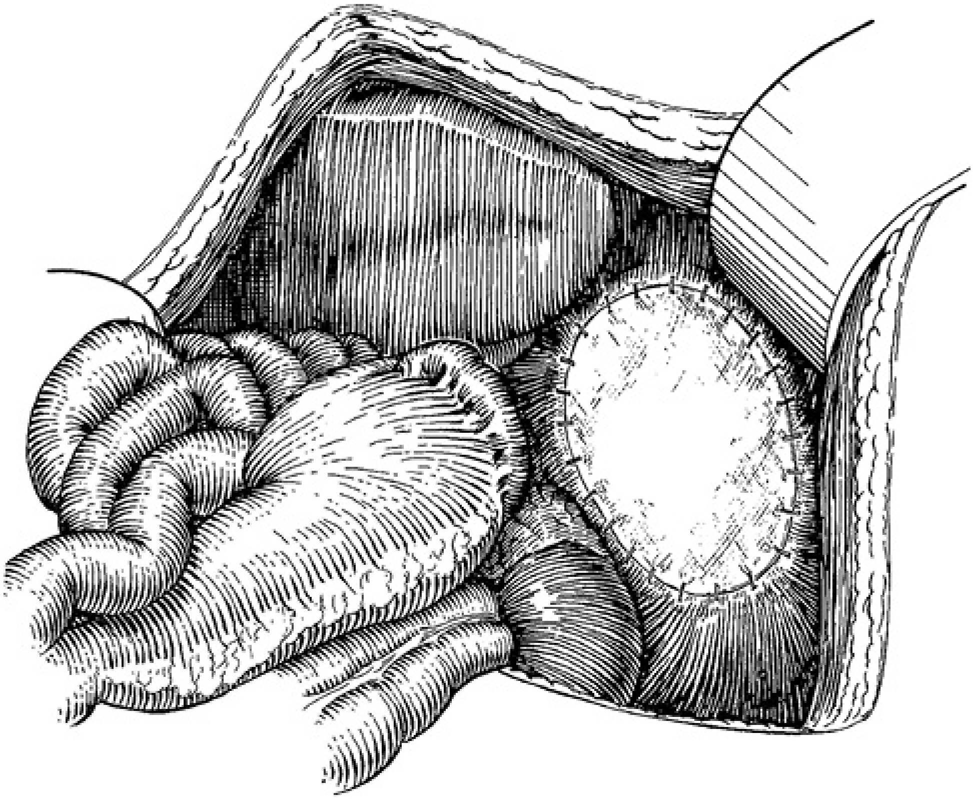


13.6a–c

If a prosthetic patch is required, 1 mm PTFE (Gore-Tex) is preferred (**Figure 13.7**). The patch should be moderately redundant and can be anchored to the ribs laterally. An ipsilateral thoracostomy tube is unnecessary unless there is concern for bleeding or an air leak.



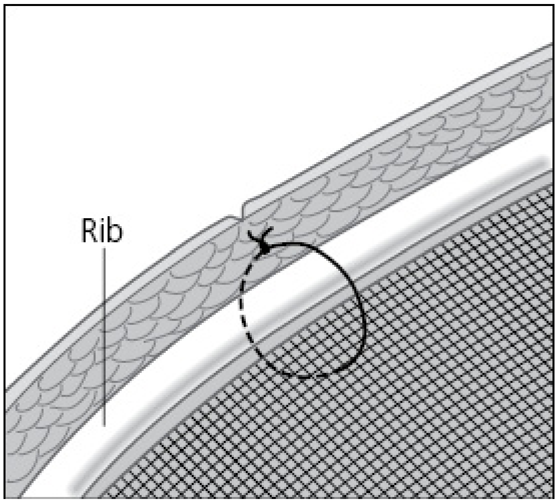
13.7

Correction of malrotated midgut or an appendectomy is not routinely indicated. Postoperative volvulus is rare, but significant bleeding after heparinization may be a major problem. Electrocautery is used as much as possible to perform the operation because of the potential need for ECMO and heparin at the time of surgery or postoperatively.

WOUND CLOSURE

Abdominal wall closure may be challenging due to insufficient abdominal domain. If the abdomen cannot be closed safely without compromising the viscera and venous return, a 1 mm PTFE patch can augment the abdominal wall temporarily. If more domain is required, a temporary silo can be employed.

In cases of true diaphragmatic agenesis, the posterior and medial rim of diaphragm usually used to anchor mesh for a patch repair are absent. A larger prosthetic patch should be used for these defects and can be placed using pericostal sutures to bring the patch to the posterior wall, through the chest wall, through the diaphragm, and back out of the chest (**Figure 13.8**). The knot is secured in the subcutaneous tissue. Recurrent diaphragmatic hernias are common in these patients.



13.8

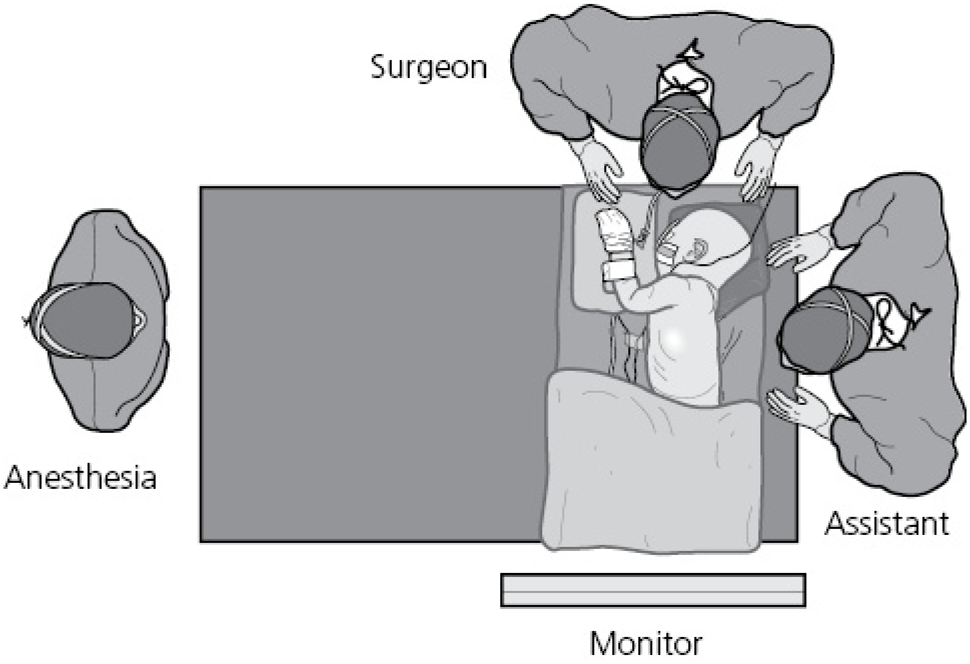
Transthoracic open repair

While a thoracotomy may be used for CDH repair, it is generally avoided due to its morbidity. Nonetheless, diaphragmatic repair through a right thoracotomy may be of value in cases of hepatopulmonary fusion or other anatomical variations that make reduction of the liver complicated. Thoracoscopic repair is also an option in stable infants. Thoracoscopic repair should not be performed on infants on ECMO due to the high risk of bleeding.

Thoracoscopic repair

POSITION OF PATIENT

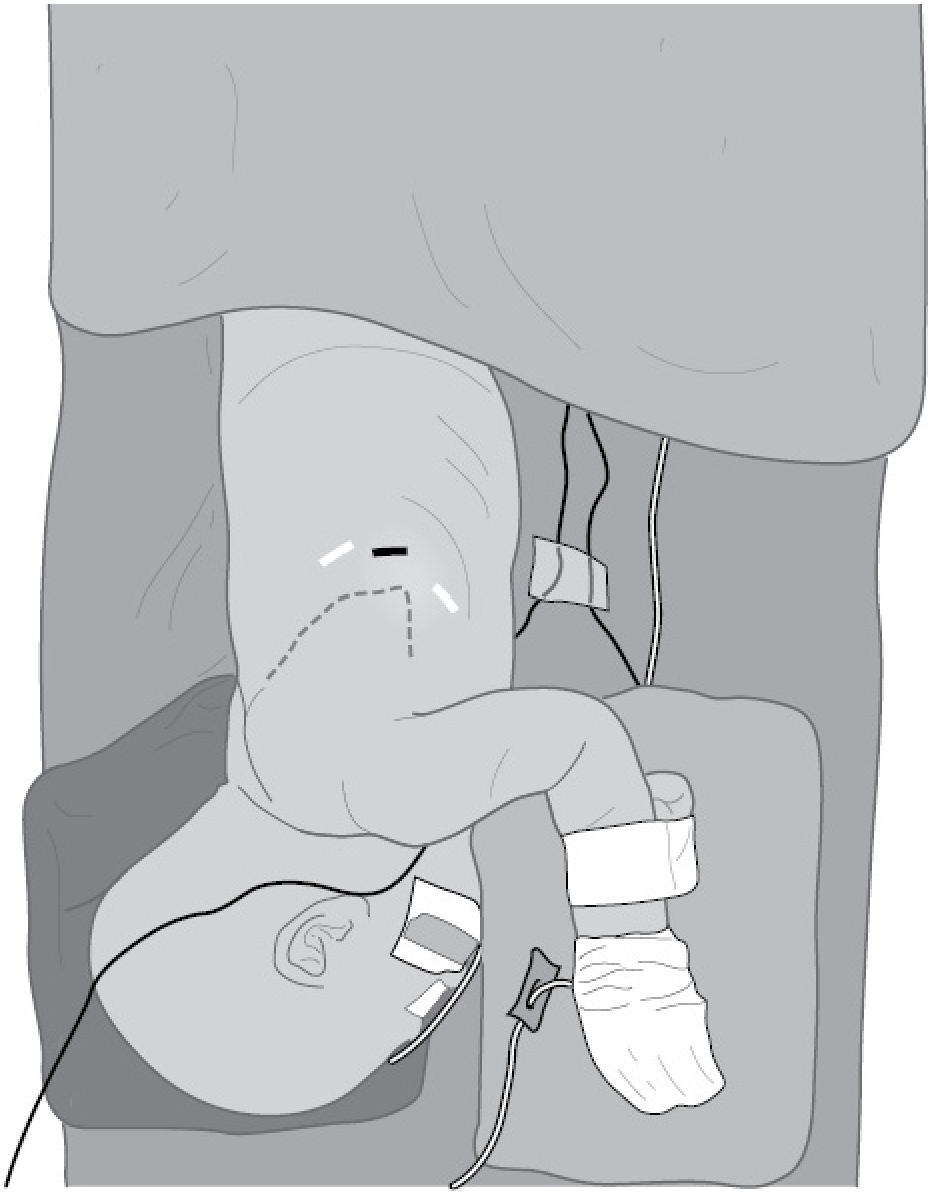
The infant is placed in a near-lateral decubitus position with a small raised support placed beneath (**Figure 13.9**). The chest is prepared from sternum to spine, including the abdomen, in anticipation of conversion to a transabdominal approach. Anesthesia should be positioned at the side of the operative table, and the operating surgeon stands at the head of the bed with a monitor by the patient's feet.



13.9

INCISION/ACCESS

A Veress needle is placed below the scapular tip in the mid-axillary line while holding respiration (**Figure 13.10**). Carbon dioxide is insufflated, achieving an intrathoracic pressure of 3–5 mmHg; this pressure can be decreased once abdominal contents are reduced in order to decrease potential respiratory acidosis. The Veress needle is then exchanged for a 5 mm trocar, and a 5 mm, 30-degree scope is inserted. Two additional 3–5 mm trocars are placed under direct visualization in the anterior and posterior axillary lines, cephalad to the camera port.

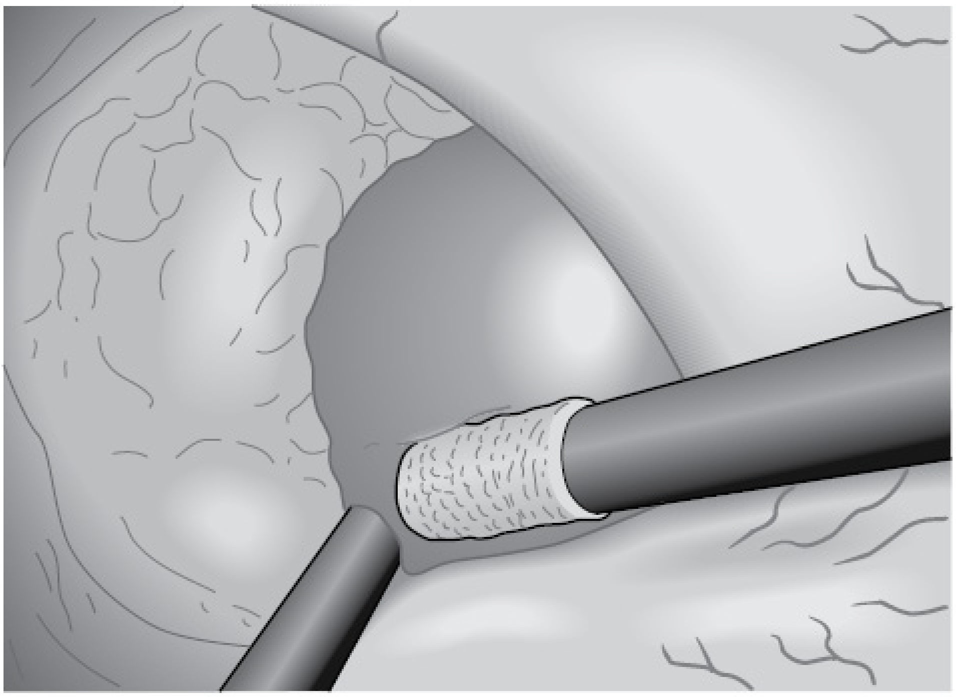


13.10

STEPS

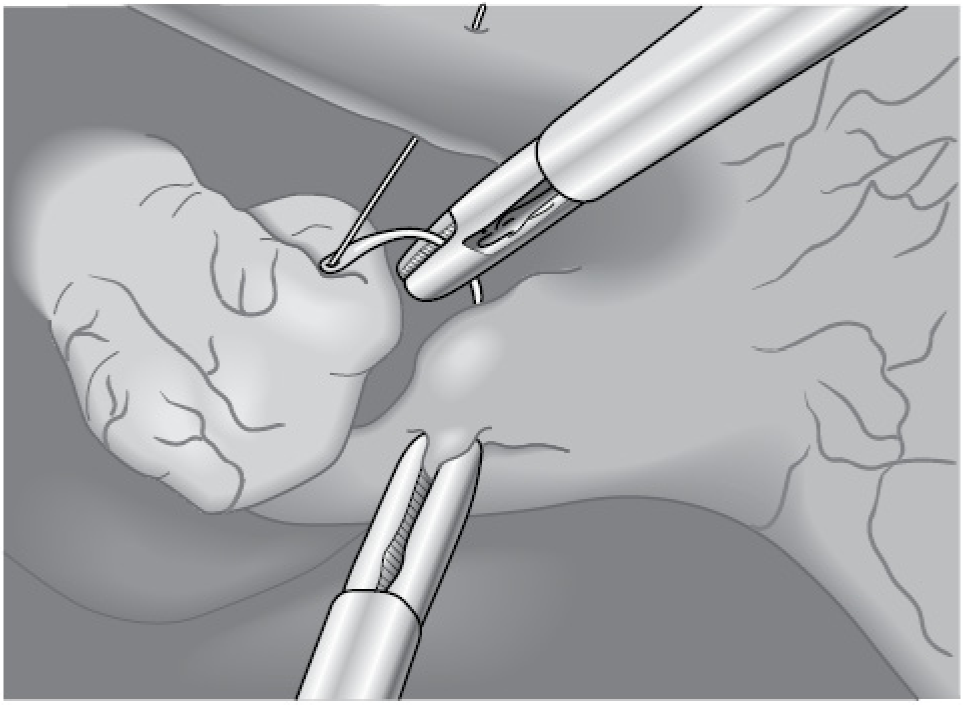
Herniated abdominal contents are reduced into the abdominal cavity.

If the diaphragmatic defect can be easily approximated, primary repair is performed using interrupted, non-absorbable suture (**Figure 13.11**). Many surgeons advocate for the use of pledgets in areas of attenuated diaphragm and cauterize the defect edge. An underlay patch may be useful in A and B defects that can be primarily repaired.



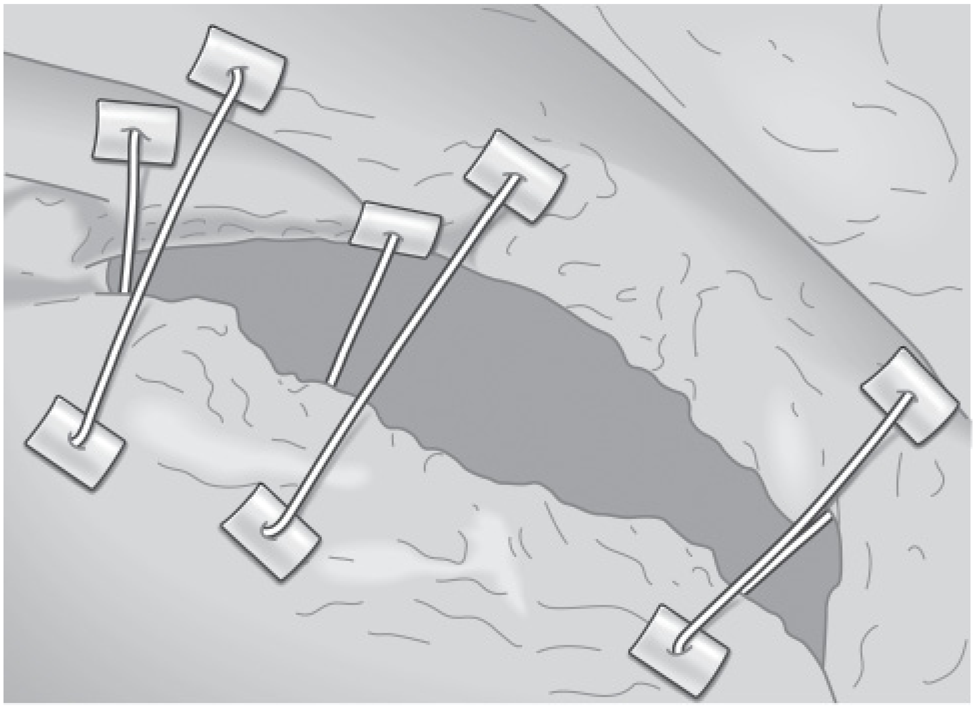
13.11

Ethibond® sutures with pledgets are preferred (**Figure 13.12**). Pericostal sutures can facilitate lateral closure of the defect as well.



13.12

External pressure on the chest wall aids approximation of diaphragmatic edges (**Figure 13.13**). When the defect cannot be closed primarily, a PTFE patch can be cut to size. Total diaphragmatic agenesis usually warrants conversion to an open transabdominal approach.



13.13

WOUND CLOSURE

The muscle is approximated with 4/0 Vicryl®, and the skin is closed with 5/0 Monocryl®. Evacuation of the pneumothorax is accomplished by inserting a catheter into the chest before closure of the final port incision. A thoracostomy tube should only be used in cases with a concern for bleeding or an air leak.

POSTOPERATIVE CARE

The therapeutic strategy used in the preoperative period is reinstituted in the postoperative period. Muscle paralysis is discontinued to allow spontaneous respiration. Adequate analgesia with narcotics is mandatory. Sufficient IV fluids are given to maintain adequate circulating blood volume and hemoglobin for oxygen delivery.

OUTCOME

These children are at risk for pulmonary, gastrointestinal, and neurologic sequelae. As such, these patients require long-term follow-up care by a multidisciplinary team consisting of pediatric surgeons, pulmonologists, cardiologists, physical and occupational therapists, and dieticians.

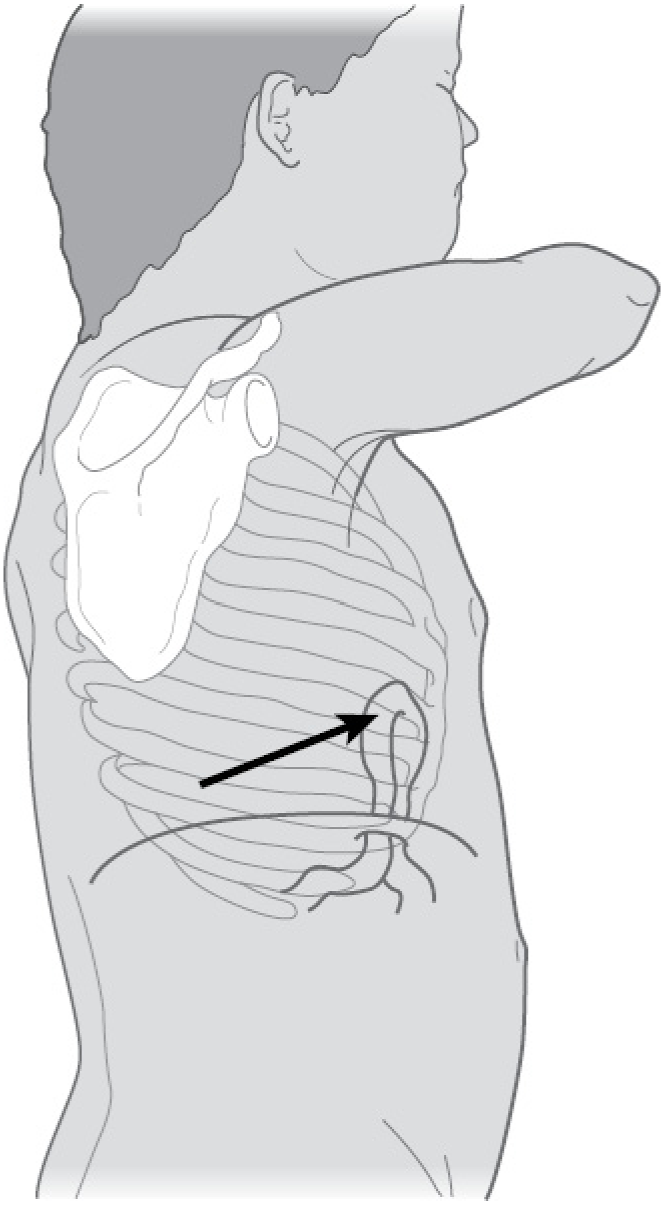
There is a risk of recurrent herniation in all patients. The risk is increased with patch repair. There is also reported increased risk of recurrence with thoracoscopic repair and some would advocate for an open repair during reoperation.

ECMO

Although this therapy is discussed in detail in a separate chapter (Chapter 14), there are some special considerations for ECMO in these patients. Because ECMO may be appropriate both before and after operation, these infants should be transferred to an ECMO center as soon as possible after diagnosis and the infant (or fetus) is stable enough for transport. When patients are repaired on ECMO, they should receive Amicar, keep platelets >100 000, and keep lower activated clotting times (ACTs). Meticulous surgical technique is essential to avoid bleeding.

MORGAGNI HERNIA

A much less common type of diaphragmatic hernia, referred to as a Morgagni hernia, occurs anteromedially on either side of the junction of the septum transversum and the thoracic wall (**Figure 13.14**). Morgagni hernias differ significantly from Bochdalek hernias in incidence, severity, presentation, and treatment.



13.14

Anterior diaphragmatic hernias account for less than 5% of all congenital diaphragmatic defects. Right-sided Morgagni hernias (90%) are significantly more common than left, due to the additional strength of the pericardial attachment to the diaphragm on the left. Bilateral Morgagni hernias are not an uncommon finding, and occasionally communicate in the midline, constituting a large anterior diaphragmatic defect. Hernia sacs are much more common in patients with Morgagni hernia (95%) compared to

Bochdalek hernia. The transverse colon and small bowel herniate most commonly.

Unlike Bochdalek hernias, Morgagni hernias rarely present in the neonatal period. When this defect is seen in the pediatric age group, recurrent pulmonary infection is the most common complaint, while gastrointestinal symptoms are much less frequent. Diagnosis can be made with a combination of posterior–anterior and lateral chest radiographs and upper gastrointestinal fluoroscopy.

Treatment of Morgagni hernias in asymptomatic patients is considered by some to be controversial. Repair can be performed transthoracically or transabdominally, however, transabdominal repair is advocated because it allows for repair of bilateral hernias, which are often only diagnosed intraoperatively. Repair consists of suturing the diaphragm to the underside of the posterior rectus sheath at the costal margin after reduction of the hernia. Most surgeons also advocate for resection of the sac, although this may increase the risk of pneumopericardium or pneumothorax. Patients with Morgagni hernias are typically more stable preoperatively than patients with Bochdalek hernia and are better candidates for minimally invasive approaches to repair.

FURTHER READING

Al-Salem AH. Congenital hernia of Morgagni in infants and children. *J Pediatr Surg* 2007; 42: 1539–43.

Boloker J, Bateman DA, Wung J-T, Stolar CJH. Congenital diaphragmatic hernia in 120 infants treated consecutively with permissive hypercapnea, spontaneous respiration, elective surgery. *J Pediatr Surg* 2002; 37: 357–66.

Burgos CM, Frenckner B, Luco M, et al. Prenatally versus postnatally diagnosed congenital diaphragmatic hernia – Side, stage, and outcome. *J Pediatr Surg* 2019; 54(4): 651–5.

Gander JW, Fisher JC, Gross ER, et al. Early recurrence of congenital diaphragmatic hernia is higher after thoracoscopic than open repair: A single institutional study. *J Pediatr Surg* 2011; 46: 1303–8.

Geggel RL, Murphy JD, Langleben D, et al. Congenital diaphragmatic hernia: Arterial structural changes and persistent pulmonary hypertension after surgical repair. *J Pediatr* 1985; 107: 457–64.

Lansdale N, Alam S, Losty PD, Jesudason EC. Neonatal endosurgical congenital diaphragmatic hernia repair: A systematic review and meta-analysis. *Ann Surg* 2010; 252: 20–26.

Putnam LR, Harting MT, Tsao K, et al. Congenital diaphragmatic hernia defect size and infant morbidity at discharge. *Pediatrics* 2016; 138(5).

Reid L. Embryology of the lung. In: de Reuck AVS, Porter R (eds). *Development of the Lung*. London: Churchill; 1967: pp. 109–30.

Silen ML, Canvasser DA, Kurkchubasche AG, et al. Video-assisted thoracic surgical repair of a foramen of Bochdalek hernia. *Ann Thorac Surg* 1995; 60: 448–50.

Simpson JS, Gossage JD. Use of abdominal wall muscle flap in repair of large congenital diaphragmatic hernia. *J Pediatr Surg* 1971; 6: 42–4.

The Congenital Diaphragmatic Hernia Study Group. Congenital diaphragmatic hernia: Defect size correlates with developmental defect. *J Pediatr Surg* 2013; 48: 1177–82.

14

Extracorporeal membrane oxygenation

THOMAS PRANIKOFF AND RONALD B. HIRSCHL

HISTORY

Extracorporeal membrane oxygenation (ECMO) has been used to describe a method of extracorporeal life support (ECLS) using extrathoracic cannulation for cardiopulmonary support. ECLS is a supportive rather than a therapeutic intervention. It provides adequate perfusion and gas exchange (venoarterial bypass) or gas exchange alone (venovenous support), and so avoids deleterious effects from high oxygen concentration and positive pressure ventilation while allowing resolution of reversible heart and lung pathology.

PRINCIPLES AND JUSTIFICATION

Vascular access for ECLS in the neonate is particularly challenging due to the small vessel size. The route of access depends on the method used. Venoarterial (VA) bypass is indicated if both cardiac and pulmonary support are required, and in neonates where access for venovenous (VV) support cannot be obtained.

VV support is the method of choice for pulmonary failure and can adequately support most infants, including those with depressed cardiac function from high pressure ventilation used to manage their severe respiratory failure prior to ECMO. VV support is more physiologic than VA and provides well-oxygenated blood to the pulmonary circulation which acts as a potent vasodilator to reduce right to left shunting. It also obviates the need for arterial cannulation and thereby lowers the risk of arterial embolization and carotid ligation or repair. Various access sites, including the umbilical, femoral and carotid/jugular vessels, have been used.

For VA access, the preferred site is the right atrium via the right internal jugular vein for venous drainage and the aortic arch via the right common carotid artery for arterial infusion. The internal jugular vein and carotid artery are relatively large in the neonate and may be distally ligated without major sequelae.

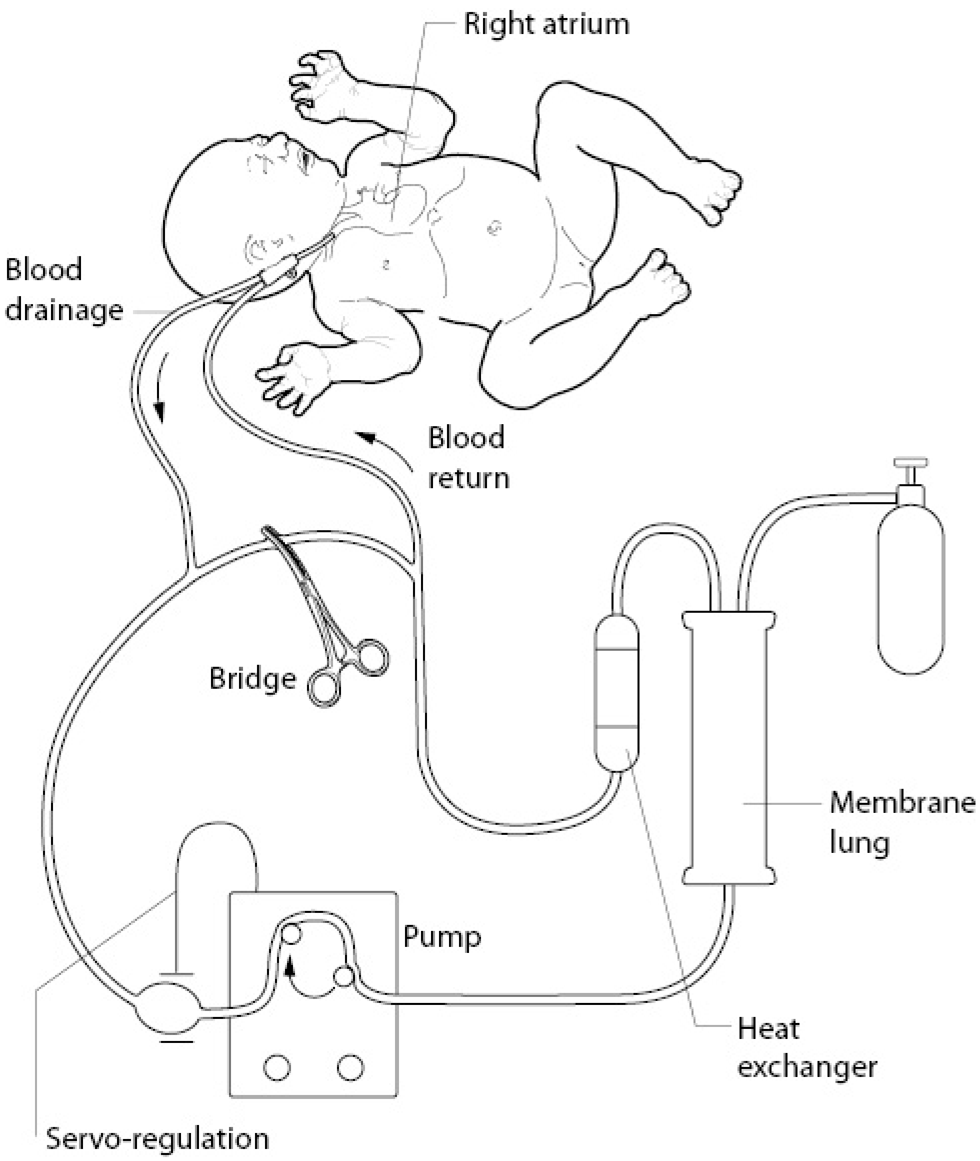
For VV access, a double lumen cannula is placed into the right atrium via the right internal jugular vein. This technique is limited by the size of the vein, because the smallest cannula currently available is 13 Fr.

Selection of technique

VA bypass requires arterial ligation to prevent distal embolization from flow past the cannula. VV support can be performed either using this technique or without vessel ligation via a percutaneous or semi-open technique. Percutaneous access utilizes the Seldinger technique to place the cannula. Because the size of the vessel in relation to the cannula is unknown, vessel disruption is a risk. Ultrasound can be used to visualize vessel size and plan for the proper size cannula. Alternatively, a semi-open technique may be used. This technique requires a small incision to visualize the size of the vein as an aid to select the correct cannula size (usually 13 Fr or 16 Fr in a newborn). Cannula insertion can also be visualized through this incision. With the percutaneous and semi-open techniques, vessel ligation is not utilized. This has several advantages: Cephalad flow into the cannula increases the amount of deoxygenated blood available to enter the bypass circuit, the vessel may remain patent after decannulation, and kinking of the cannula at the vessel is reduced.

ECMO circuit

Blood is drained from the right atrium via the venous cannula into a small bladder by siphon/gravity. As long as venous drainage is adequate to fill the bladder, blood enters the raceway (tubing within the pump head) where it is actively pumped into the membrane lung. Here, blood travels in a countercurrent fashion to the sweep gas, separated by a thin silicone membrane. Oxygen enters the blood and carbon dioxide enters the sweep gas by simple diffusion along a concentration gradient. Blood then enters the heat exchanger where it is warmed to body temperature before entering the arterial cannula. This blood may enter either the arterial (VA bypass) or the venous system (VV support) (**Figure 14.1**).


Right atrium
Blood drainage
Blood return
Bridge
Membrane lung
Pump
Heat exchanger
Servo-regulation

14.1

PREOPERATIVE

Preparation

Vascular cannulation and decannulation are performed in the neonatal intensive care unit under adequate sedation and neuromuscular blockade. Neuromuscular blockade is especially important in preventing the potentially lethal complication of an air embolus during introduction of the venous cannula. The instruments and sterile procedures used are identical to those used in the operating room. Heparin sodium (100 units/kg) is drawn up for subsequent administration.

Anesthesia

Local anesthesia is administered by infiltration of 1% lidocaine (lignocaine).

OPERATIONS

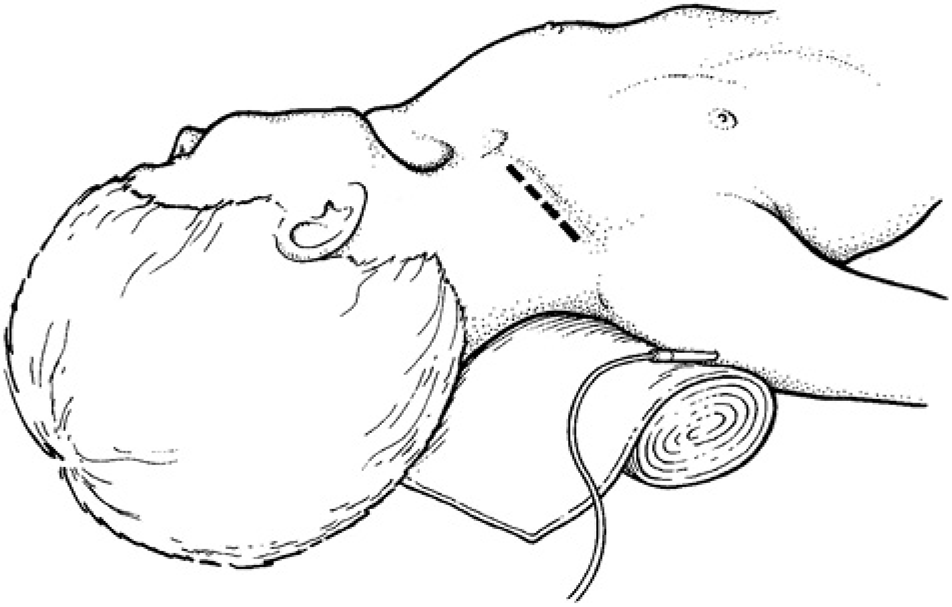
Position of patient

The patient is placed supine with the head turned to the left. A roll is placed transversely beneath the shoulders. Special attention is paid to assuring that the endotracheal tube is positioned to prevent kinking under the drapes during the procedure. This can be accomplished by using a piece of suction tubing split lengthwise and placed over the tube at the connector to prevent kinking. The chest, neck, and right side of the face are aseptically prepared and draped.

Venoarterial/venovenous cannulation: Open technique

INCISION

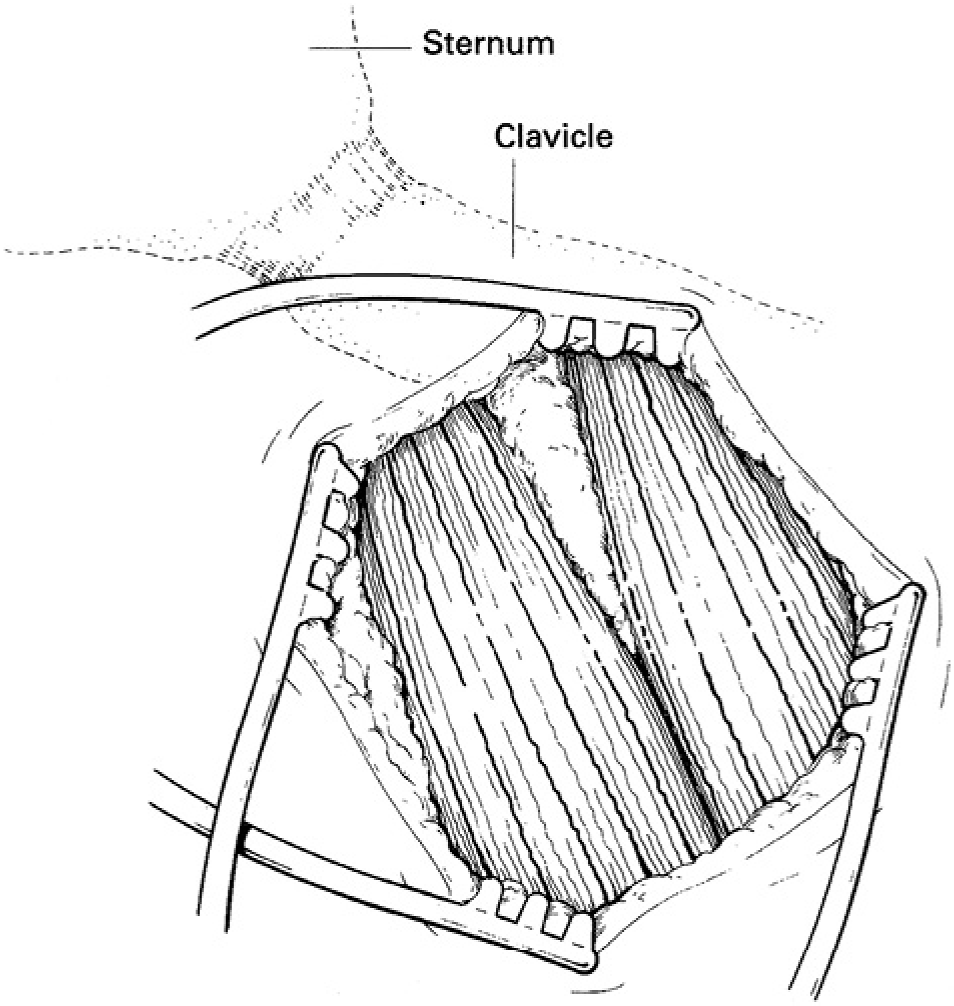
A transverse cervical incision approximately 2–3 cm in length is made one finger's breadth above the clavicle over the lower aspect of the right sternocleidomastoid muscle (**Figure 14.2**).



14.2

EXPOSURE OF THE CAROTID SHEATH

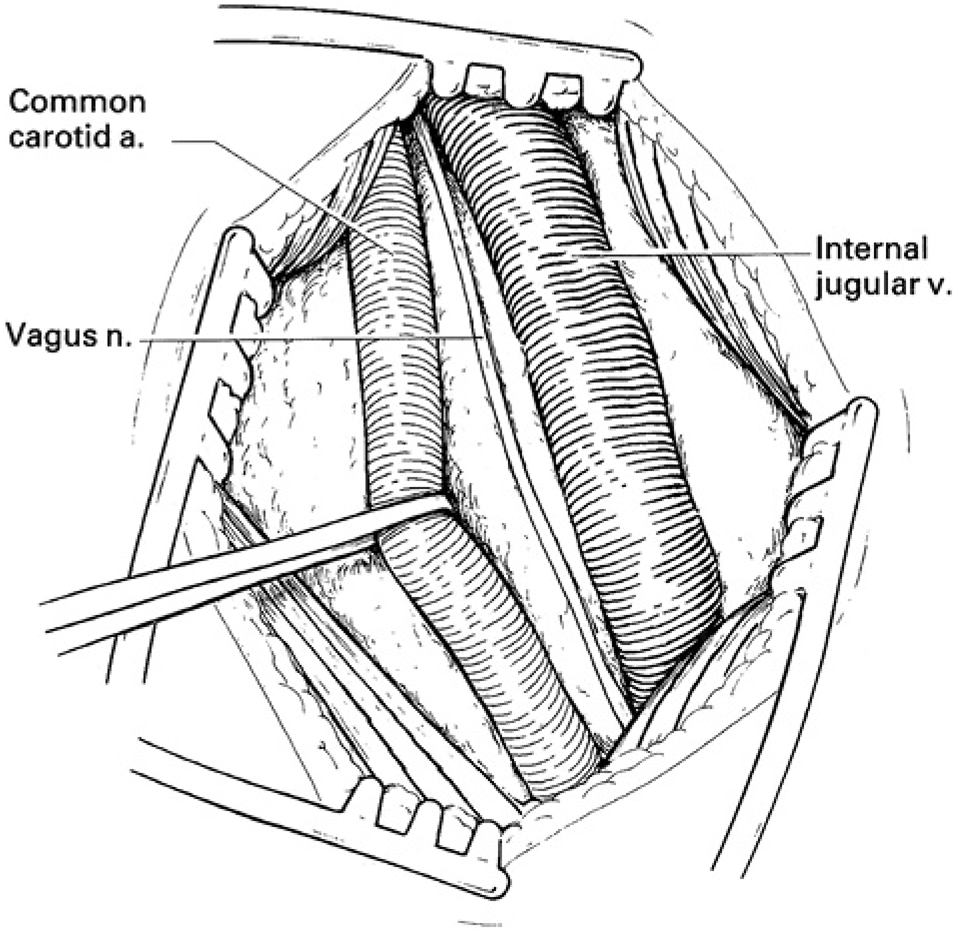
The platysma muscle and subcutaneous tissues are divided with electrocautery and the sternocleidomastoid muscle is exposed. Dissection is continued bluntly between the sternal and clavicular heads of the muscle. The omohyoid muscle will be seen superiorly. It may be necessary to divide the omohyoid muscle tendon to expose the carotid sheath. Two alternating self-retaining retractors are placed (**Figure 14.3**).



14.3

DISSECTION OF THE VESSELS

The carotid sheath is opened and the internal jugular vein, common carotid artery, and vagus nerve are identified and isolated (**Figure 14.4**). Dissection is progressed proximally and distally along the vessels, dissecting the vein first. Special care should be taken while dissecting the vein to avoid induction of spasm, which makes subsequent introduction of a large venous cannula difficult. Manipulation of the vein therefore should be minimized. There is often a branch on the medial aspect of the internal jugular vein which must be ligated. Ligatures of 2/0 silk are placed proximally and distally around the internal jugular vein. The common carotid artery lies medial and posterior and has no branches, which makes its dissection proximally and distally safe. Ligatures of 2/0 silk are also placed around the carotid artery. The vagus nerve should be identified.

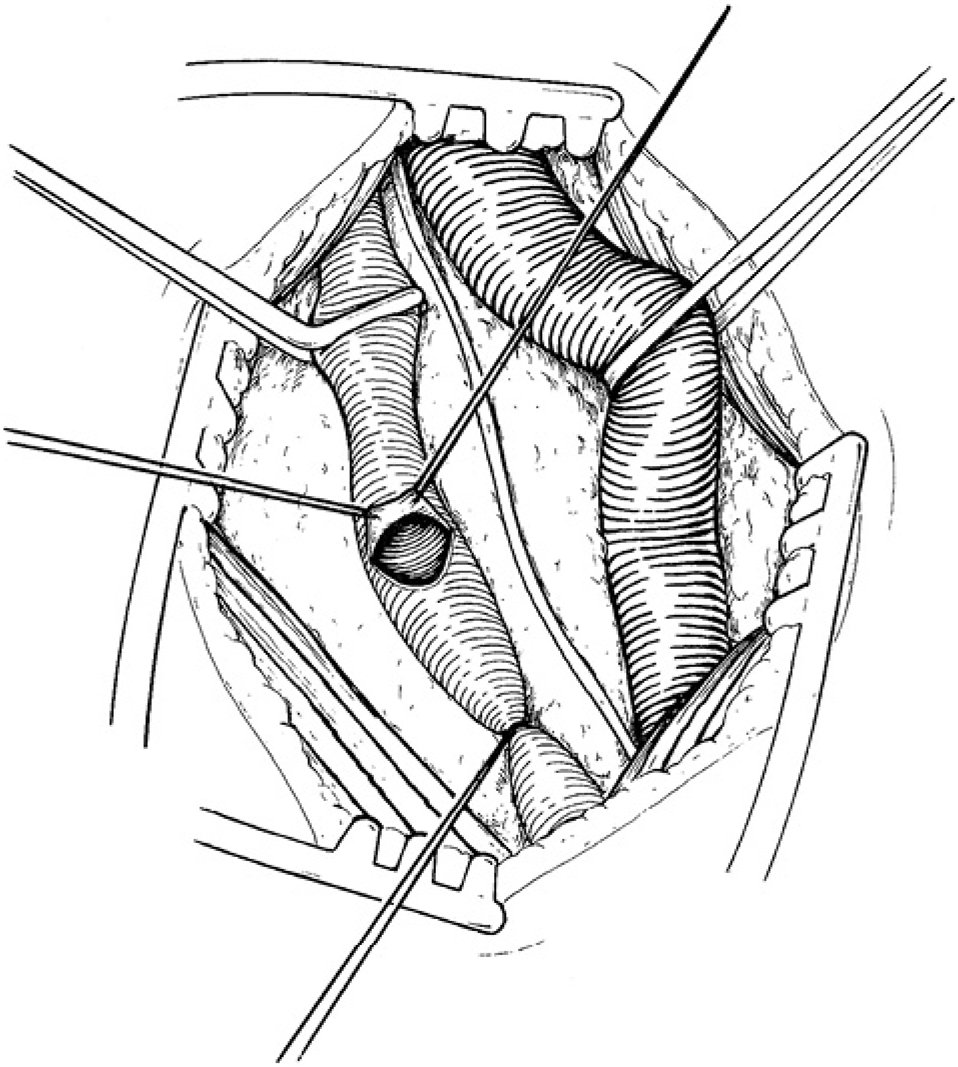


14.4

Once vessel dissection is completed, heparin (100 units/kg) is administered intravenously and 3 minutes allowed for circulation. During this waiting period, papaverine is instilled into the wound to enhance vein dilatation.

ARTERIOTOMY/VENOTOMY

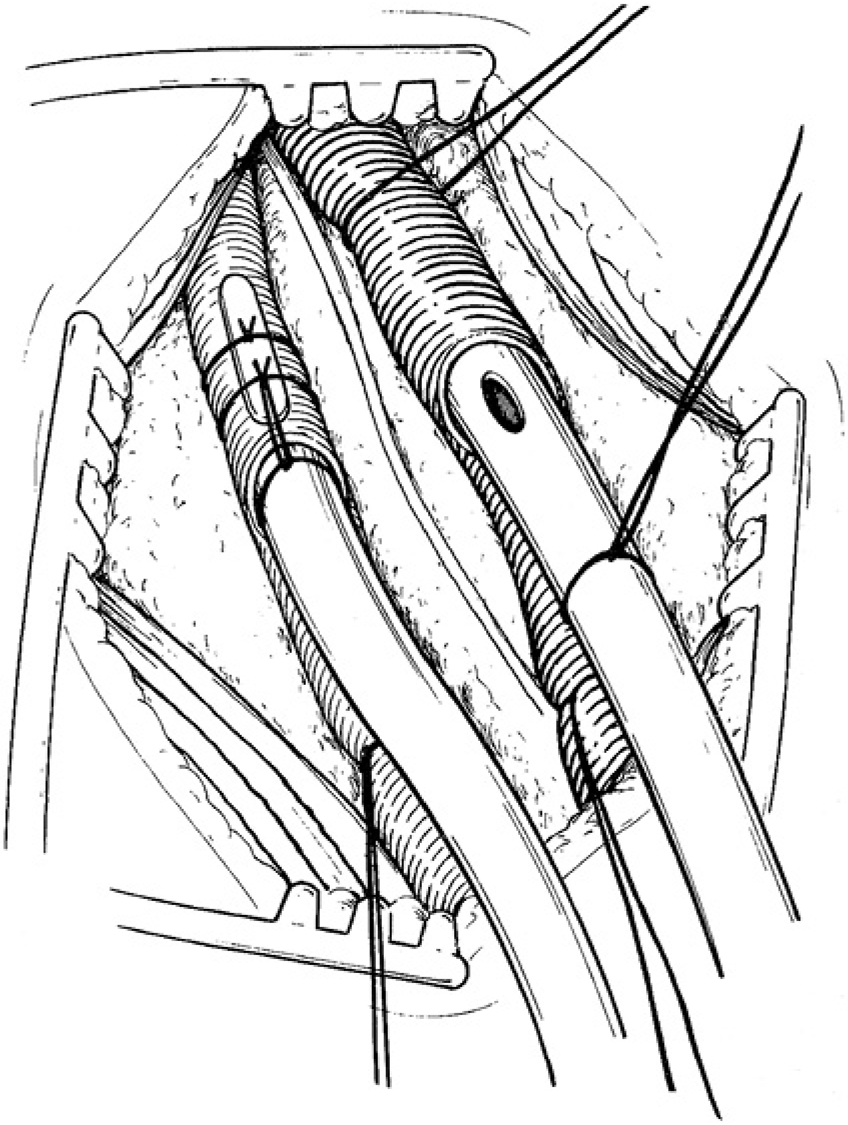
For venoarterial bypass, the arterial cannula is chosen (usually 10 Fr) and marked with a 2/0 silk ligature, left uncut, at a point that will allow the tip of the cannula to lie at the ostium of the brachiocephalic artery (about 2.5 cm). The venous cannula (usually 12–14 Fr) is similarly marked at a point equal to the distance from the venotomy to the right atrium (roughly 6 cm). An obturator is placed into the venous cannula to prevent blood from flowing out through the side holes during introduction into the vessel. The common carotid artery is ligated distally (**Figure 14.5**). Proximal control is obtained with the use of an angled ductus clamp. A transverse arteriotomy is made near the distal ligature. Full-thickness stay sutures of 6/0 polypropylene are placed on the proximal edge of the artery to prevent subintimal dissection during cannula insertion. Following arterial cannulation, a venotomy is performed in similar fashion. Gentle retraction of the caudal ligature around the vein precludes the need for a ductus clamp during venotomy and venous cannulation. Stay sutures can also aid venous cannulation but can be used selectively.



14.5

CANNULA PLACEMENT

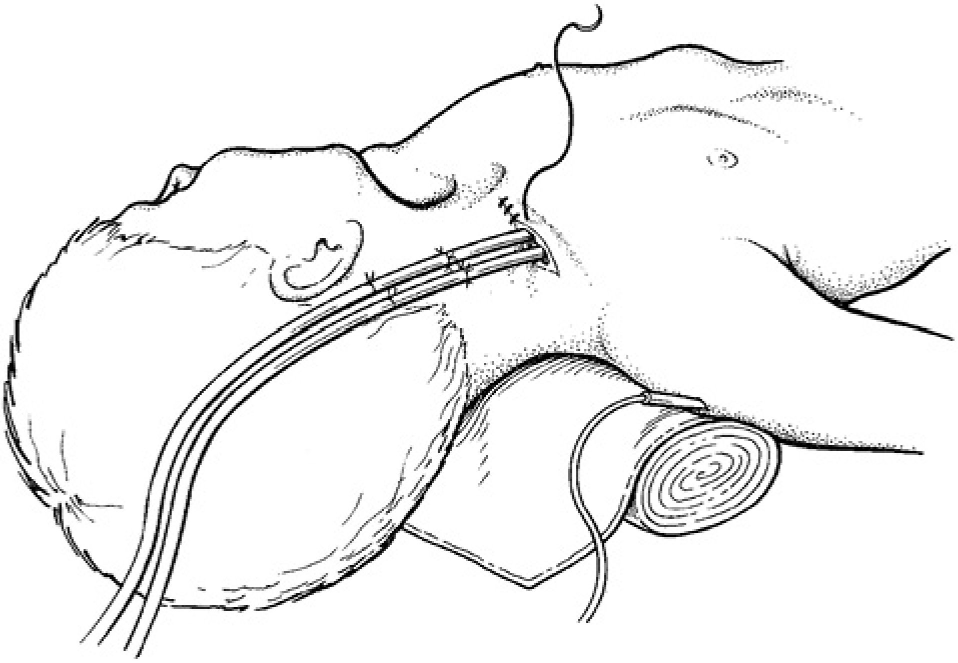
The cannulas are carefully placed into the artery and vein and secured using two circumferential 2/0 silk ligatures, with a small piece of Silastic vessel loop inside the ligatures to protect the vessels from injury during decannulation when the ligatures are sharply divided. The ends of the marking ligatures are tied to the most distal circumferential ligature for extra security (**Figure 14.6**). Immediately after each cannula is secured, it is carefully debubbled via back-bleeding and filling with heparinized saline. For venovenous support, the double-lumen cannula is placed into the venotomy and advanced 5.5 cm. It is crucial to maintain the arterial reinfusion (red) port anteriorly while securing for proper orientation to minimize recirculation of reinfused blood.



14.6

WOUND CLOSURE

The wound is irrigated with saline and hemostasis obtained. The skin is closed with continuous monofilament suture (**Figure 14.7**). The wound is dressed with gauze. The cannula is sutured to the skin with several 2/0 silk sutures. Special attention should be directed to affixing the cannulas securely to the bed.

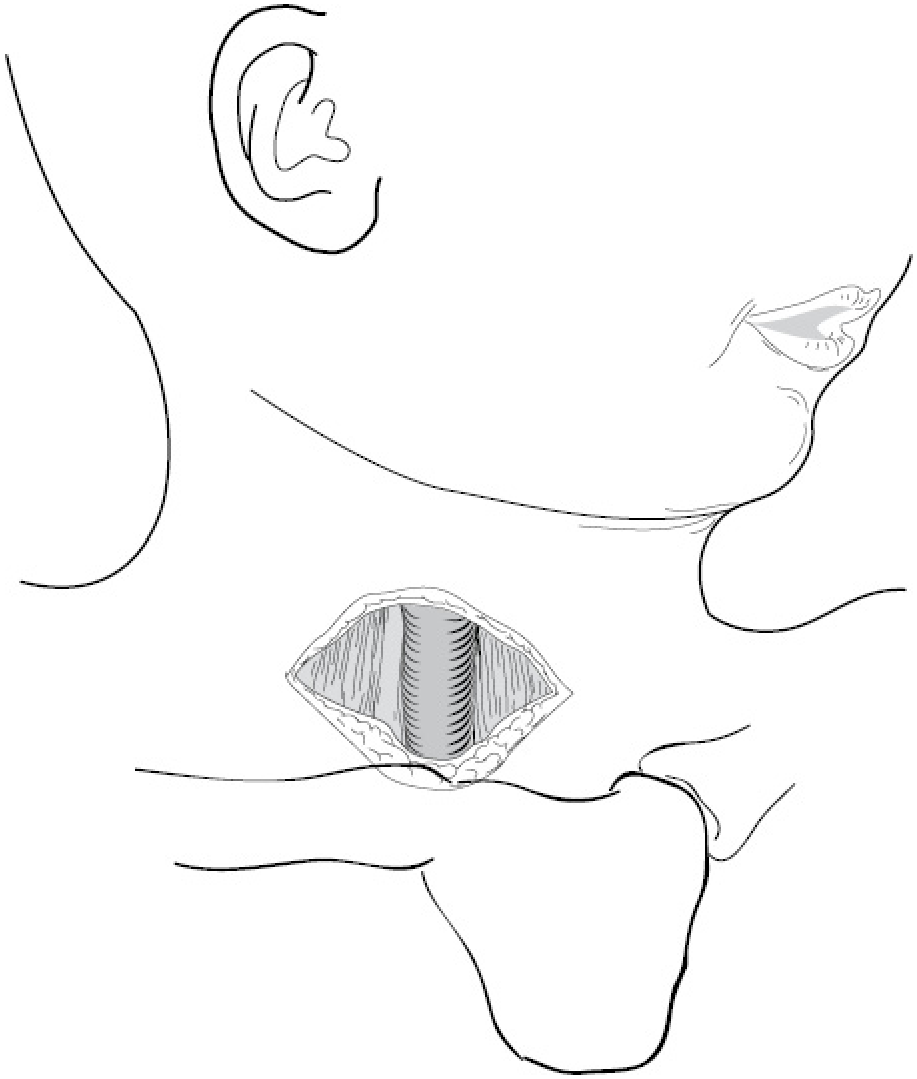


14.7

Venovenous cannulation: Semi-open technique

INCISION AND VEIN EXPOSURE

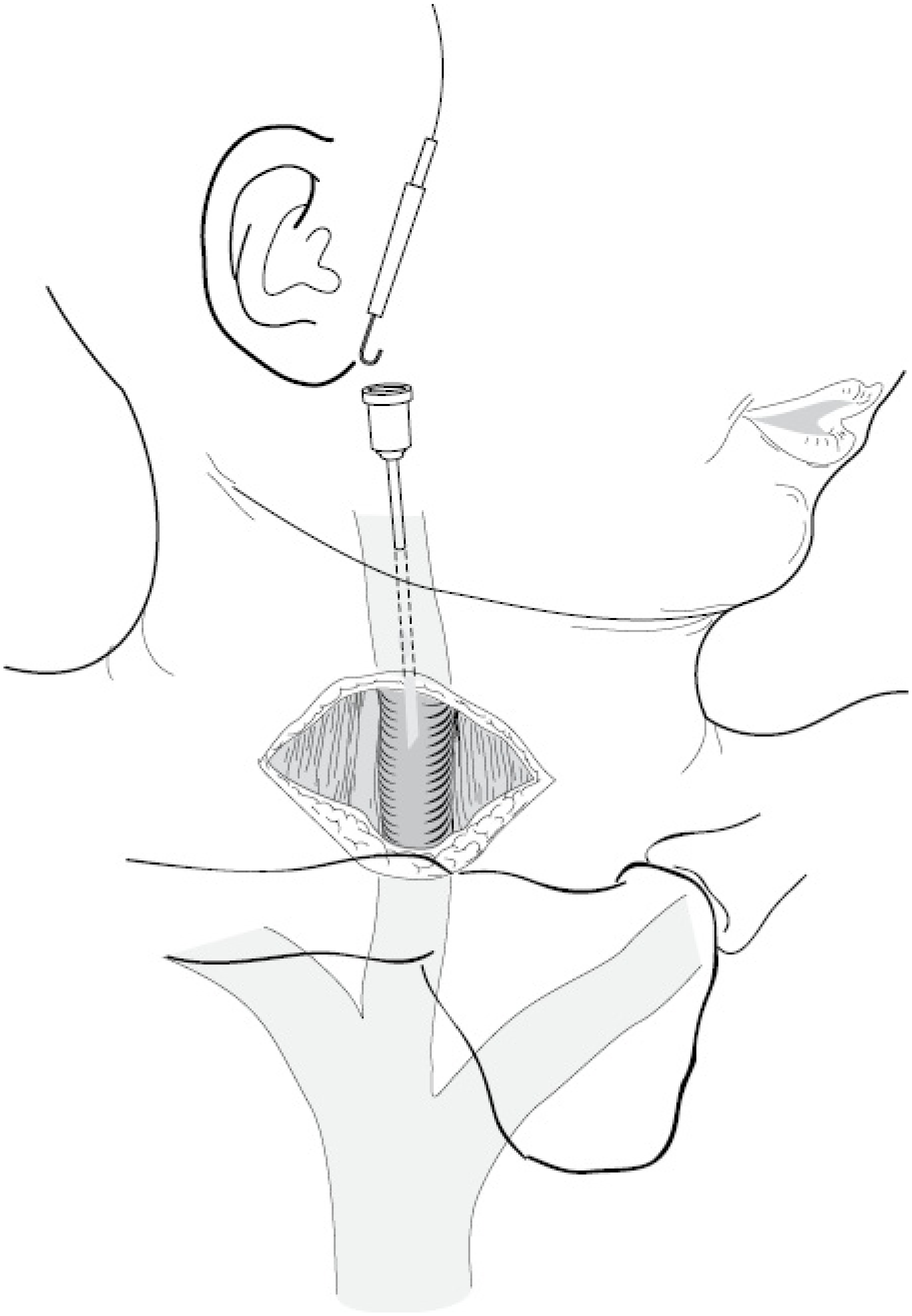
A transverse cervical incision approximately 1.5–2 cm in length is made 2 cm above the right clavicle between the heads of the sternocleidomastoid muscle. The platysma is divided with electrocautery and the anterior surface of the internal jugular vein is exposed with minimal dissection (**Figure 14.8**). The vessel is observed and either a 12 Fr or a 15 Fr Origen venovenous ECMO cannula (Origen Biomedical Inc., Austin, TX) is selected.



14.8

GUIDEWIRE PLACEMENT

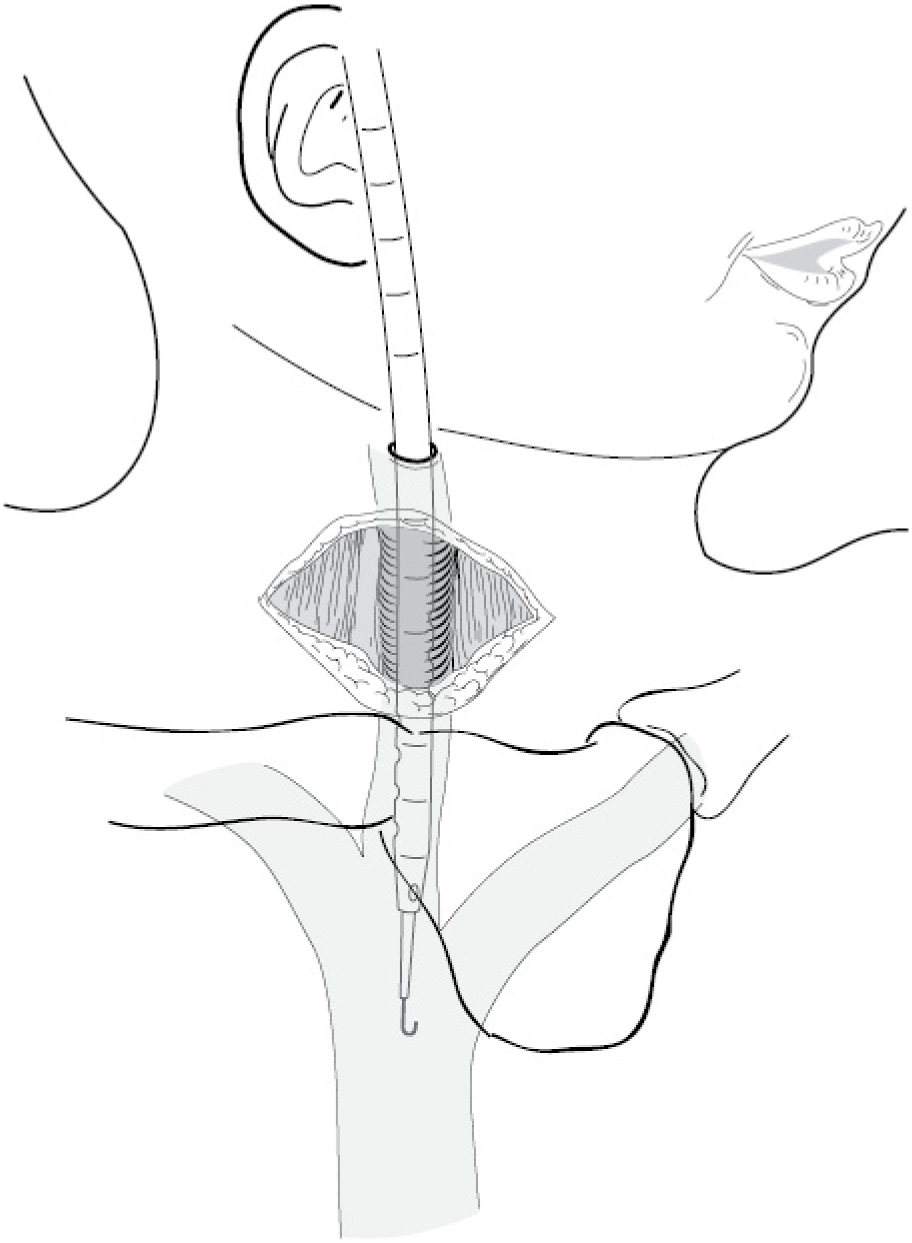
The cannula skin exit position is selected so that the cannula will lie behind the right ear when the head is returned to the midline (**Figure 14.9**). The needle/catheter is placed through the skin 2 cm superior to the incision and into the internal jugular vein to enter either under the skin flap or just inside the incision. The needle is removed and a 0.89 mm (0.035 inch) diameter guidewire is advanced and the catheter is withdrawn. A Teflon guiding obturator is placed over the guidewire into the vessel and right atrium. The use of fluoroscopy, if available, is helpful. The skin exit is slightly enlarged with a scalpel.



14.9

CANNULA PLACEMENT

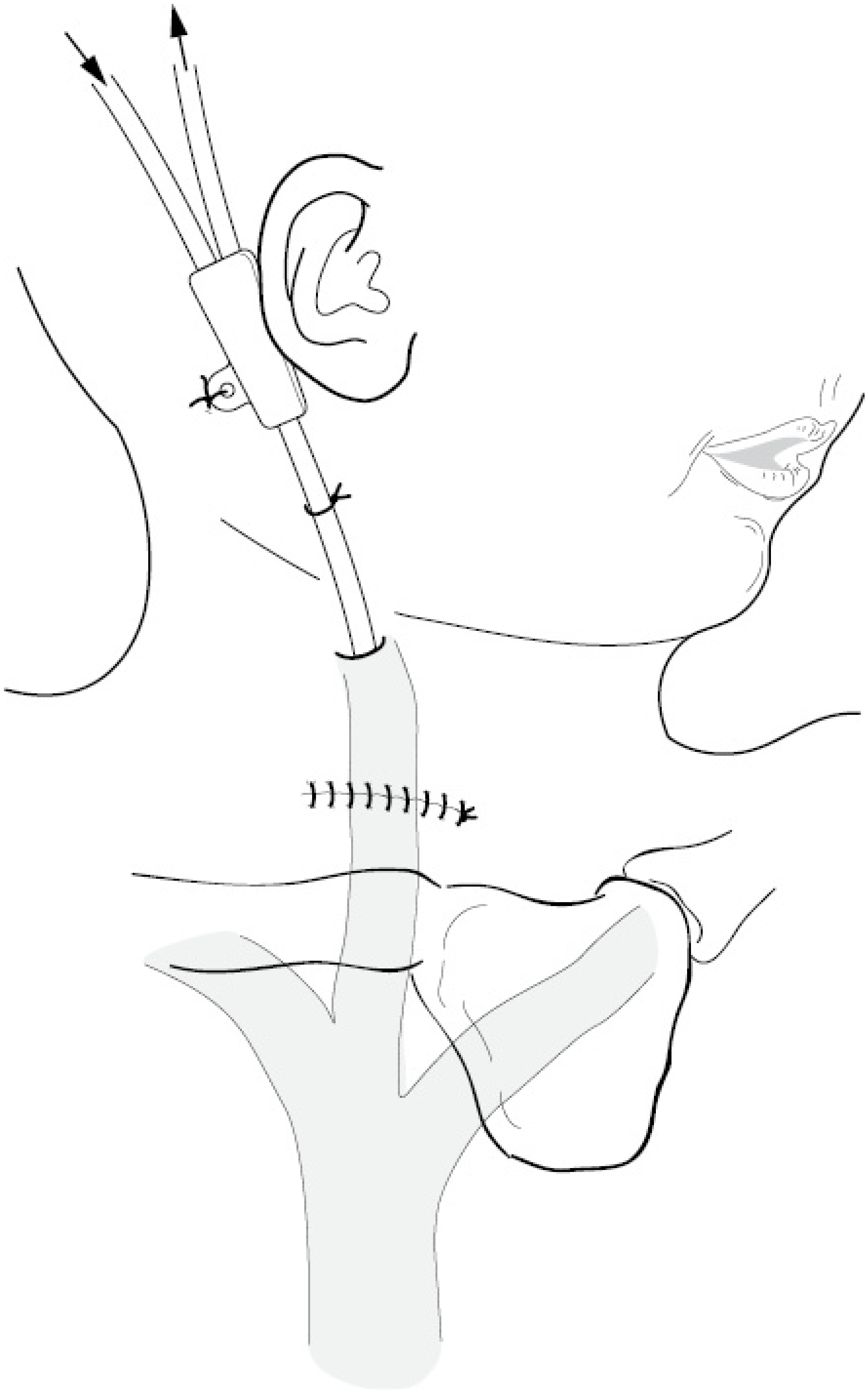
Heparin (100 units/kg) is administered and 3 minutes allowed for circulation. The selected cannula is advanced over the Teflon obturator into the vein under direct vision to confirm entrance into the vein (**Figure 14.10**). The arterial (red) port of the cannula must be directed anteriorly to allow the arterial blood to cross the tricuspid valve and minimize recirculation of circuit blood. The tip of the cannula is placed at 6–9 cm from the skin.



14.10

WOUND CLOSURE AND CANNULA FIXATION

The relatively low venous pressure allows adequate hemostasis around the venotomy site without any ligature. This prevents kinking of the thin-walled cannula, which often occurs at the area of a ligature if used around the vessel. Repositioning of the cannula only requires removing the skin sutures, repositioning the cannula, and replacing skin sutures. The cannula is fixed to the skin with several 2/0 silk sutures. The incision is closed with a monofilament suture (**Figure 14.11**).



14.11

DECANNULATION

After respiratory failure has resolved to allow ventilation without extracorporeal support, decannulation can be performed by removing the skin sutures, pulling the cannula, and holding pressure on the catheter exit site for 5 minutes or until bleeding stops. Alternatively, a purse-string suture can be placed around the cannulation site and tied after cannula removal. Care must be taken to rapidly remove the entire cannula to prevent air from entering the side holes while the end of the cannula remains in the vessel.

Venovenous cannulation: Percutaneous technique

CANNULA SELECTION AND VEIN ACCESS

Ultrasound is used to evaluate the right internal jugular vein for patency and size. It is critical to be sure the vein is large enough to accommodate the cannula selected to prevent vein injury. The diameter in millimeters multiplied by three will yield the French size. The vein is accessed 2 cm superior to the clavicle under direct ultrasound visualization with a micropuncture needle (5 Fr micropuncture access kit, Cook Medical, Bloomington, IN). A flexible 0.45 mm (0.018 inch) × 40 cm (10 inch) guidewire with flexible tip is advanced into the right atrium using fluoroscopy. If guidewire placement is unsuccessful, conversion to a semi-open or open technique should be considered. A 5 Fr coaxial catheter/dilator is advanced over the guidewire and the inner cannula removed. This is then replaced with a 0.89 mm (0.035 inch) J-tip guidewire and either a 13 Fr or a 16 Fr Origen venovenous ECMO cannula (Origen Biomedical Inc., Austin, TX) is selected.

CANNULA PLACEMENT

Heparin (100 units/kg) is administered and 3 minutes allowed for circulation. The skin exit is slightly enlarged with a scalpel. The tract is dilated and the cannula advanced under fluoroscopy. It is optimal to have the guidewire advanced into the inferior vena cava so that the atrium is not injured during cannula advancement. The tip of the cannula is placed at approximately 6–9 cm from the skin.

CANNULA FIXATION

The relatively low venous pressure allows adequate hemostasis around the venotomy site. If the skin exit site is not tight around the cannula, a purse-string suture provides hemostasis. Repositioning of the cannula only requires removing the skin sutures, repositioning the cannula, and replacing skin sutures. It is crucial to maintain the arterial reinfusion (red) port anteriorly while securing for proper orientation to minimize recirculation of reinfused blood. The cannula is fixed to the skin with several 2/0 silk sutures. Echocardiography may be used to assure that the jet of blood from the arterial infusion port is entering the tricuspid valve.

DECANNULATION

After respiratory failure has resolved to allow ventilation without extracorporeal support, decannulation can be performed by removing the skin sutures, pulling the cannula, and holding pressure on the catheter exit site for 5 minutes or until bleeding stops. Alternatively, a purse-string suture can be placed around the cannulation site and tied after cannula removal. Care must be taken to rapidly remove the entire cannula to prevent air from entering the side holes while the end of the cannula remains in the vessel.

POSTOPERATIVE CARE

The cannulas are connected to the extracorporeal bypass circuit, assuring that no air bubbles are present, and bypass is initiated. Dopamine infusion into the ECLS circuit reinfusion connector is often necessary for inotropic support with the initiation of venovenous support if the patient required this type of support prior to ECLS initiation. A chest radiograph should be obtained following the procedure to verify optimal cannula placement distal to the aortic arch (arterial cannula) and inferior aspect of the right atrium (venous cannula). Bleeding from the wound is controlled by lowering ACTs, platelet transfusions, administration of fresh frozen plasma and local instillation of fibrin glue. Bleeding not controlled by these maneuvers should be investigated and controlled by operative exploration after the open technique. Placement of a purse-string suture around the catheter exit site controls bleeding after the semi-open technique.

OUTCOME

More than 30,000 neonates have been treated since 1972, with an overall survival rate of 73%. The four most common diagnoses requiring ECLS in neonatal patients are meconium aspiration syndrome, congenital diaphragmatic hernia, pneumonia/sepsis, and persistent pulmonary hypertension. Survival rates of 92%, 51%, 57% and 73%, respectively, have been achieved (2013–2018).

FURTHER READING

Hogan MJ. Neonatal vascular catheters and their complications. *Radiol Clin North Am* 1999; 37(6): 1109–25.

Peek GJ, Firmin RK, Moore HM, Sosnowski AW. Cannulation of neonates for venovenous extracorporeal life support. *Ann Thorac Surg* 1996; 61: 1851–2.

Pranikoff T, Hines MH. Vascular access for extracorporeal support. In: Van Meurs K, Lally KP, Peek G, Zwischenberger JB (eds). *ECMO – Extracorporeal Cardiopulmonary Support in Critical Care* 3rd ed. Ann Arbor, MI: Extracorporeal Life Support Organization; 2005.

Reickert CA, Schreiner RJ, Bartlett RH, Hirschl RB. Percutaneous access for venovenous extracorporeal life support in neonates. *J Pediatr Surg* 1998; 33: 365–8.

15

Eventration of the diaphragm

COLIN G. DELONG, AFIF N. KULAYLAT, AND ROBERT E. CILLEY

HISTORY

Eventration of the diaphragm refers to the radiographic finding of an abnormally elevated hemidiaphragm. When this condition leads to symptomatic respiratory insufficiency, it may require operative intervention. The physiologic consequences of the loss of diaphragm function are made worse in infants due to the mobility of the mediastinum. When the normal hemidiaphragm descends during inspiration, the mediastinum shifts to the normal side while the side of the eventration paradoxically elevates. Surgical treatment (diaphragmatic plication) is based on removing the laxity of the abnormal diaphragm leaf to prevent paradoxical motion. Since its initial demonstration in adults in 1923 and children in 1946, numerous techniques of plication have proven effective including recent advances in laparoscopic/thoracoscopic (mid 1990s) and robotic approaches (early 2010s). The principle of creating a taut hemidiaphragm has remained the same. Novel approaches to eventration treatment including phrenic nerve repair and nerve grafting, prosthetic fixation of the diaphragm, and diaphragmatic pacing have been applied successfully in a small number of unique cases; however, surgical plication remains the mainstay of therapy.

PRINCIPLES AND JUSTIFICATION

Diaphragmatic eventrations are classified as either acquired (paralytic eventration) or congenital (non-paralytic eventration). Compromise of the phrenic nerve, most commonly occurring during intrathoracic surgery, is the cause of acquired eventration and represents the most common presentation of the condition to pediatric surgeons. Congenital eventration is thought to be related to abnormal migration of myoblasts during early embryologic development, though its precise cause is unknown and is likely to be multifactorial. In such cases, the normally muscular diaphragm is found to be grossly thinned and to demonstrate diaphragmatic fibrous dysplasia on histopathology. In its most extreme form, eventration is indistinguishable from a congenital diaphragmatic hernia with a hernia sac. Phrenic nerve injury from birth trauma is similar to acquired eventration in that the diaphragm is developmentally normal.

Diaphragm eventration may be discovered incidentally as an elevated hemidiaphragm on a chest radiograph, or it may be the cause of severe symptoms such as respiratory failure or pneumonia. Small eventrations with minimal compromise of lung volume may be monitored by serial radiographs. Recurrent symptoms or respiratory distress that requires mechanical ventilation are indications for surgical correction. When the possibility of reversible phrenic nerve injury exists, such as that due to birth trauma or operative injury, a period of observation is indicated in the symptomatic patient. If there is no improvement in diaphragm function, diaphragm plication is performed after a reasonable period of observation (2–4 weeks). Some function may eventually return to the previously paralyzed diaphragm, but this may require many months. Operative plication does not preclude some recovery of diaphragm function.

Numerous techniques have been successfully demonstrated for the correction of eventration. Specific abnormalities lend themselves to repair by different methods. The abnormally lax diaphragm may be removed by excision and closure of the resultant defect, suture of a portion of the diaphragm to the chest wall, or various methods of “gathering” or “pleating” the excess tissue with sutures. Each technique must create a taut diaphragm that is mechanically resistant to elevation from the intrathoracic pressure present during spontaneous breathing.

No portion of the diaphragm needs to be excised in cases of acquired eventration. The muscular diaphragm may ultimately regain some function, and excision only increases the risk of additional injury to the intradiaphragmatic portion of the phrenic nerve. In congenital eventrations with muscular aplasia or atrophy, the thinned portion of the diaphragm may be excised, allowing the edges that will be brought together to be visualized precisely. Pledgets and non-absorbable sutures provide the most secure closure. Since diaphragmatic defects come in all shapes and sizes, the precise orientation of the plication procedure and the decision to excise some or all of the eventration must be made on an individual basis.

Both open and endoscopic techniques have been utilized, with comparable durability of repair. Endoscopic techniques have recently gained favor for the majority of repairs in children, with reported improvements in postoperative pain and cosmetic results and low open conversion rates. Improvements in time to extubation and hospital length of stay have also been described. Endoscopic techniques are more technically challenging to complete, limiting their widespread use. However, newer techniques utilizing endoscopic staplers or other strategies for plication have increased the options for minimally invasive repair.

The choice of transabdominal versus transthoracic approaches depends on several clinical factors and surgeon preference. Thoracoscopic techniques allow for better visualization of the course of the phrenic nerve and afford a larger working space. Laparoscopic techniques confer the benefit of less intercostal pain and avoidance of single lung ventilation. Simultaneous operation for bilateral eventration or concurrent abdominal pathology is facilitated by this approach. For the most common presentation of eventration (isolated, unilateral), transthoracic approaches are the most widely reported.

PREOPERATIVE

Assessment and preparation

Unilateral eventration is suspected when the right hemidiaphragm is greater than two rib levels higher than the left, or when the left hemidiaphragm is more than one rib level higher than the right. A rare bilateral eventration is suspected when respiratory failure is present in association with radiographic demonstration of bilateral diaphragm elevation. Chest radiographs may be misleading in the patient on positive pressure mechanical ventilation. The most convincing diagnostic tests are those that allow dynamic visualization of diaphragm function during spontaneous respiration including fluoroscopy and ultrasonographic imaging. Absent or paradoxic elevation of the hemidiaphragm during spontaneous inspiratory effort is diagnostic.

Recent series have demonstrated associated malformations in up to 20% of congenital eventrations, with hypoplastic lung (10%) and congenital heart disease (5%) occurring most frequently. An echocardiogram should be performed in the presence of congenital eventration to rule out associated structural cardiac abnormalities. Diaphragm eventration has also been associated with intestinal rotational abnormalities and gastric volvulus in rare genetic conditions. Upper gastrointestinal (GI) contrast studies or computed tomography (CT) scanning may be indicated if associated malformations are suspected.

Anesthesia

General endotracheal anesthesia is used. Unilateral intubation improves exposure, but, if difficult to perform, or not well tolerated, it is not mandatory because the lung is easily retracted. Low pressure CO_2 insufflation facilitates the thoracoscopic approach. Adequate levels of insufflation during plication keep the grasped portion of the redundant diaphragm collapsed, preventing inadvertent injury to abdominal organs. Intraoperative orogastric intubation with regular gastric aspiration is important since a dilated stomach is at risk of injury when diaphragmatic sutures are passed.

OPERATIONS

Whatever technique is chosen for repair, it is imperative that a taut diaphragm is created and that adjacent organs are not injured. The stomach, colon, and spleen are at particular risk during repair of left-sided eventration via a thoracic approach.

For illustrative purposes, the diagrams below refer to the open repair of a left-sided acquired eventration with a structurally normal but non-functioning hemidiaphragm.

In the case of a congenital eventration, the central portion of the diaphragm is “thinned out.” Although the diaphragm may be “gathered” (see the section ‘Transthoracic repair of diaphragm eventration’) in a fashion similar to the repair of an acquired eventration, excising the thin central portion of the diaphragm allows the edges to be visualized clearly so that sutures may be placed in normal muscularized tissue and the abdominal viscera avoided. If excessive tension is required to bring the tissues together, a prosthetic patch may be used to close the defect. This is rarely necessary.

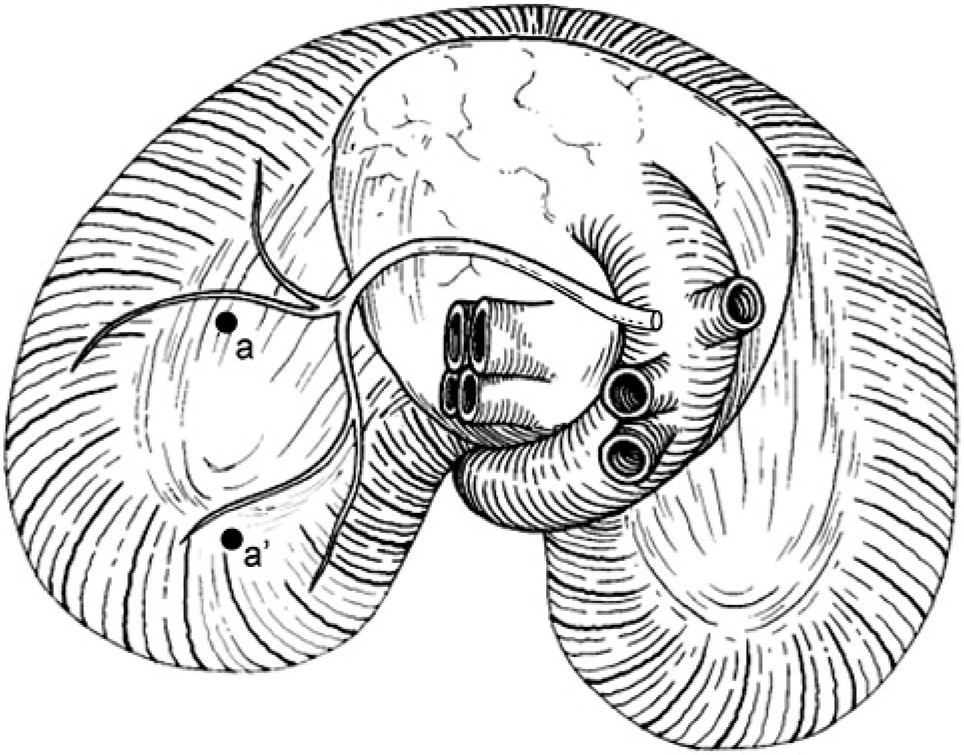
In the open repair, a transverse upper abdominal incision is used for bilateral eventration or in cases of unilateral eventration with malrotation. A lateral muscle-sparing seventh intercostal space thoracotomy is used for the open repair of an isolated left or right eventration.

Thoracoscopic procedures use three or four ports. Single port thoracoscopic plication has been demonstrated in adult patients but not yet in children. Patients are placed in the lateral position, with the camera port entering superiorly. Two working ports are then triangulated to allow anterior–posterior orientation of the repair. A fourth port is placed to assist with the reduction of the abdominal viscera if needed. Laparoscopic procedures typically use three ports with the child in supine position.

There is no consensus regarding the orientation of the plication. Anterior–posterior, right–left, and a combination of the two have been reported. Additionally, numerous variants of placing the plicating sutures have been effectively utilized, ranging from a single large fold (“reefing the mainsail") to numerous small folds ("pleating") of the redundant diaphragmatic tissue.

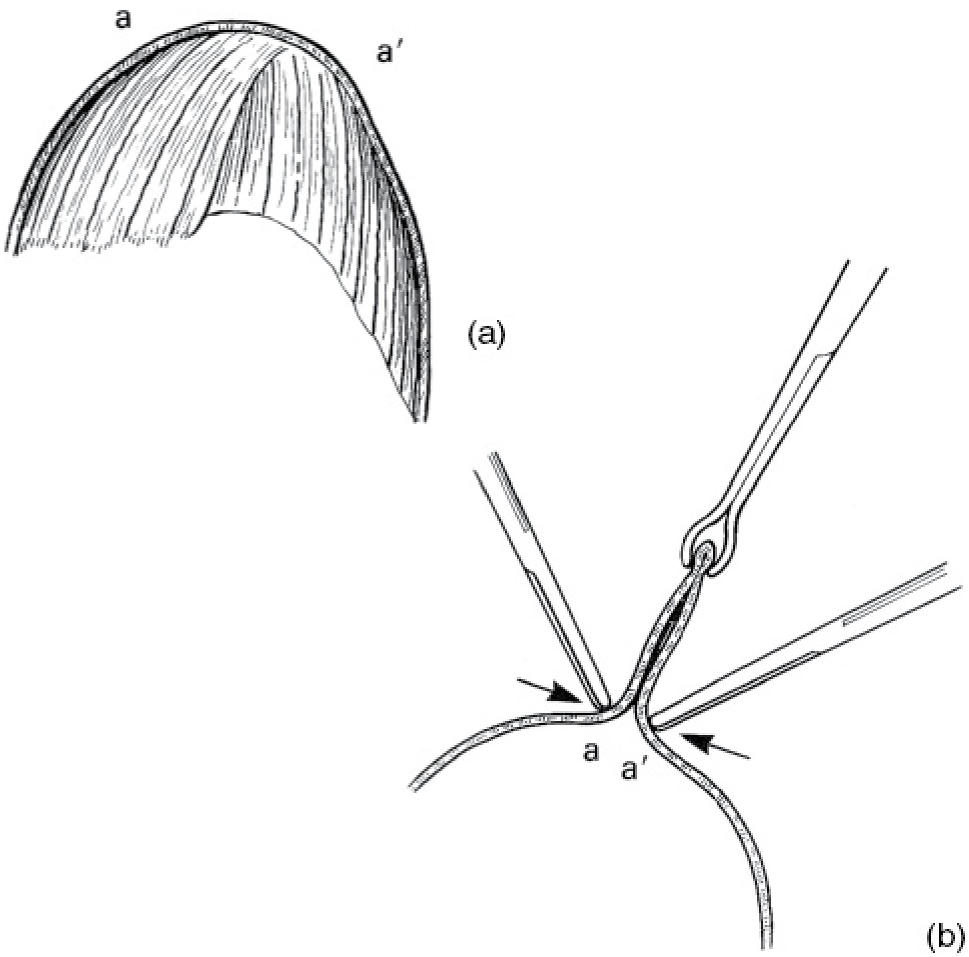
Transthoracic repair of diaphragm eventration

The main phrenic nerve on each side divides into an anterior and posterior division. Subsequent divisions usually include a sternal branch immediately off the anterior division and a bifurcation of the posterior division. The branches run in a medial to lateral orientation, allowing sutures to be placed to minimize the risk of injury to the muscular braches of the nerve. Viewed from above in **Figure 15.1**, the points a and a' represent the portions of the diaphragm that will be brought together by the plication.



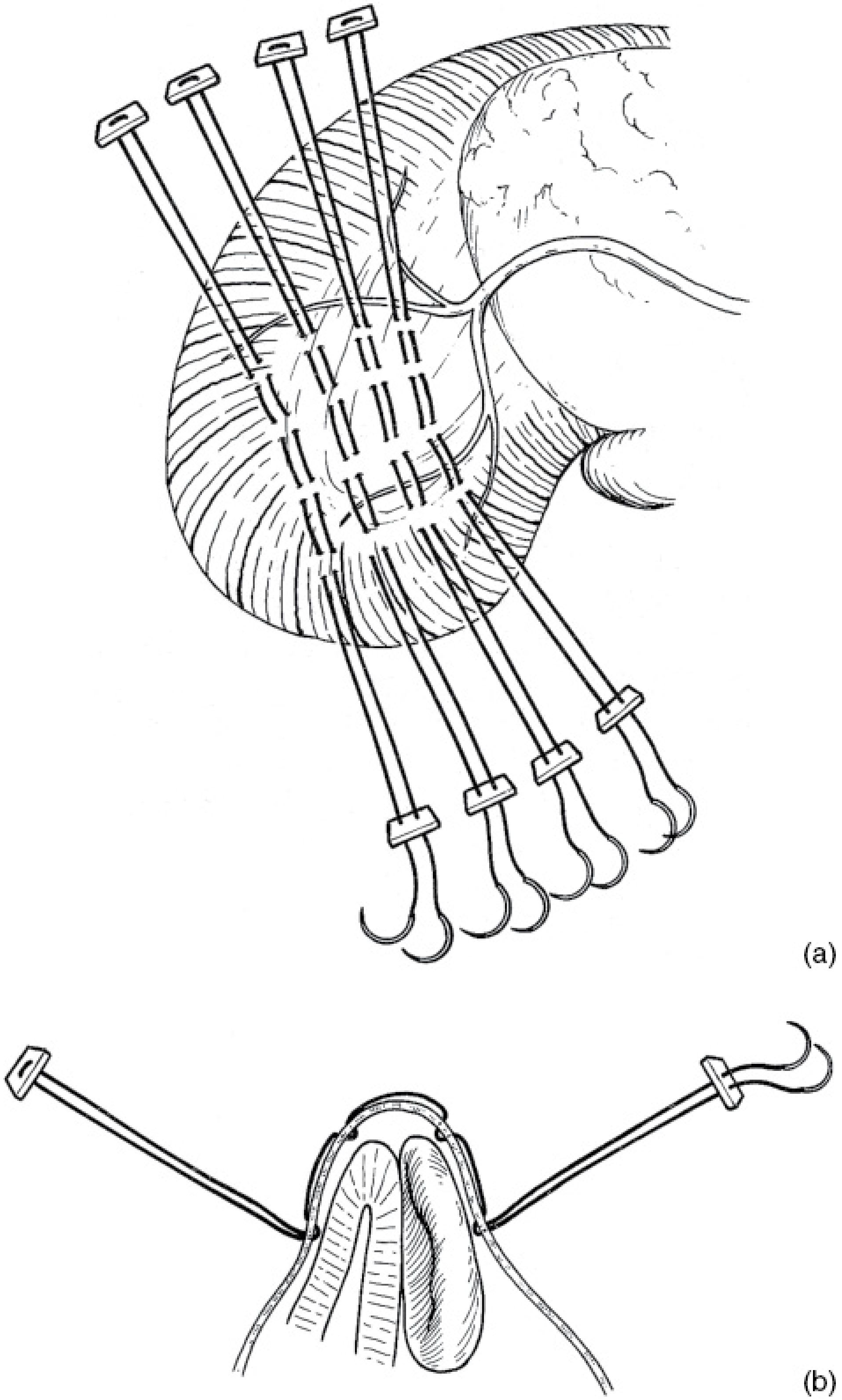
15.1

The diaphragm is grasped and manipulated to determine the amount that must be included in the plication to create a taut closure (**Figure 15.2a**). This is conveniently performed by grasping the center of the diaphragm with a non-crushing clamp. The extent of the plication, determined by manipulations with the two forceps, is marked in **Figure 15.2b**.



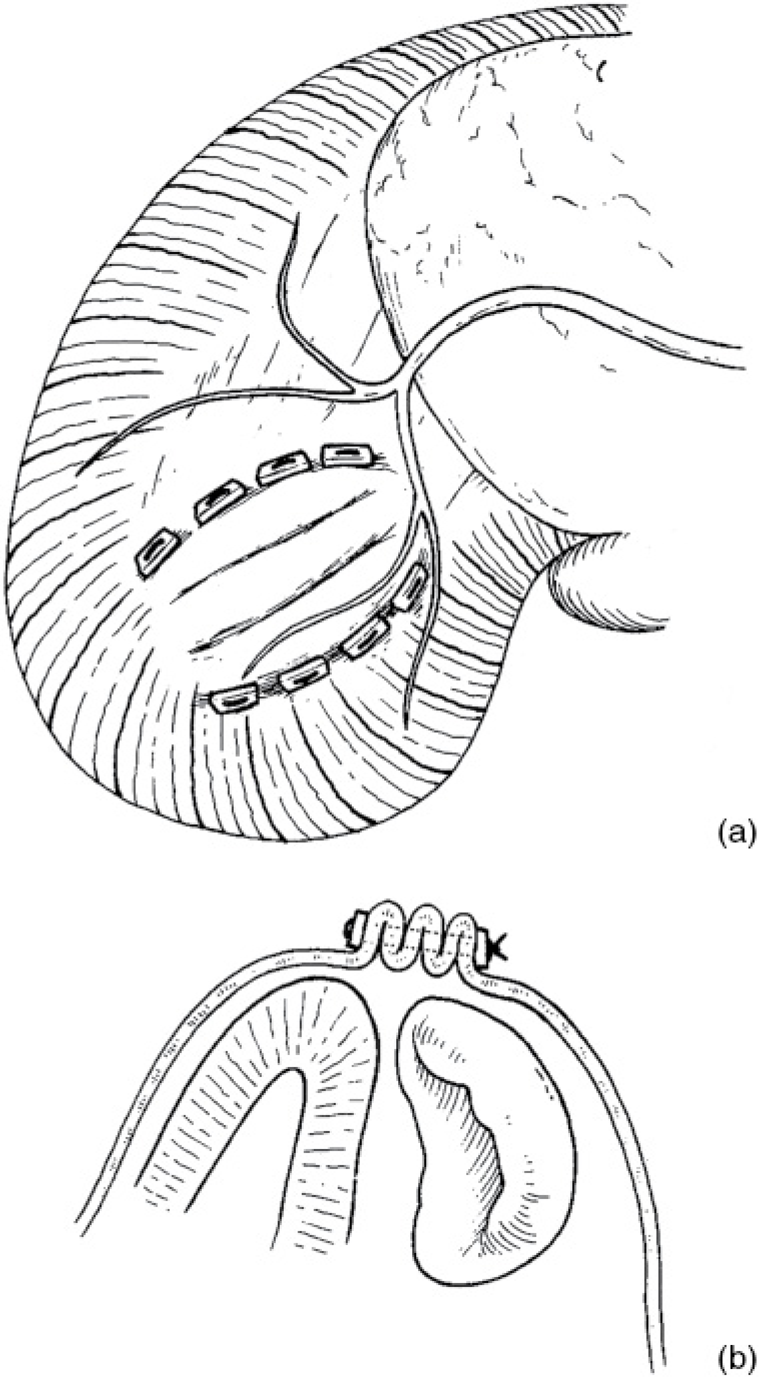
15.2a,b

Several non-absorbable sutures are placed to bring the marked portions of the diaphragm together. The sutures are passed through the intervening diaphragm muscle three or four times (**Figure 15.3a**). This maneuver is referred to as “gathering,” “reefing,” or “pleating.” Pledgeted mattress sutures are shown. Care must be taken that the sutures are passed adequately through muscle, but not deeply enough to penetrate adjacent abdominal viscera (**Figure 15.3b**).



15.3a,b

The final result of the plication creates a taut diaphragm (**Figure 15.4a and b**).

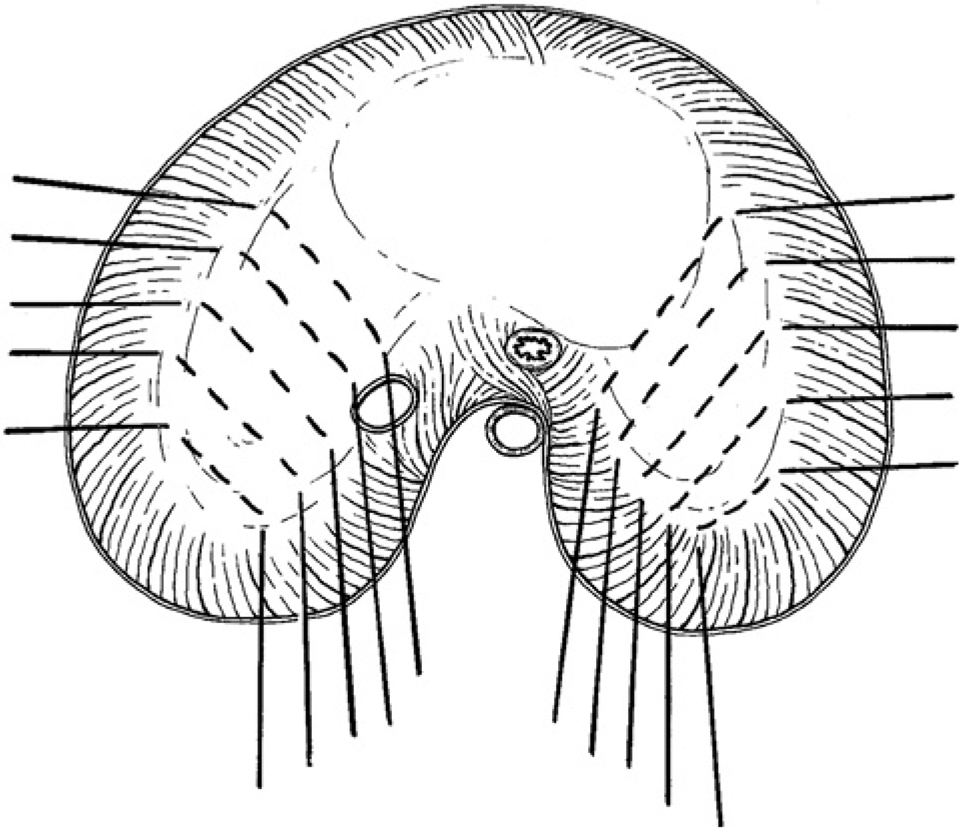


15.4a,b

Transabdominal repair of bilateral eventration

Rotational abnormalities of the intestine are addressed if present.

The undersurface of the diaphragm is exposed on the left by mobilizing the liver as necessary. The stomach and spleen are retracted and mobilized to give complete exposure. The right lobe of the liver is mobilized if needed. Plicating sutures are arranged to avoid the phrenic nerve based on its expected location (**Figure 15.5**).



15.5

POSTOPERATIVE CARE

Response to operation may be immediate, allowing prompt weaning and extubation. In patients who have required prolonged mechanical ventilation prior to operation, slower ventilator weaning is performed. The most common postoperative complications include pneumonia and pleural effusion. Intrapleural drainage may be used briefly after surgery and is usually discontinued within a few days.

OUTCOME

Although death may result from chronic respiratory failure and pneumonia, outcome is largely dependent on the presence of associated conditions, such as pulmonary hypoplasia or congenital heart disease. Long-term survival is variably reported as 69–100%. Children with bilateral eventration fare less well.

Surgical correction is durable and recurrence requiring repeat plication is rare. Inadequate plication may result if the diaphragm is not made taut at the initial operation, and the plication process will need to be repeated.

Transdiaphragmatic injury may occur with either open or thoracoscopic techniques. Intra-abdominal injuries, such as gastric or colon perforation, require immediate operative attention. Herniation into the site of the repair can occur. Rare cases of incarcerated diaphragmatic hernia and diaphragmatic rupture have been reported and should be considered in the event of severe clinical decompensation.

Many patients examined years after diaphragm plication have evidence of appropriate, although diminished, movement of the involved side with significant and stable improvement in pulmonary function tests.

Endoscopic approaches have rapidly gained popularity over open techniques of diaphragm plication. No studies have proven significant long-term differences in outcomes between the two approaches. Small technical differences, such as the ability to assess the completeness of the plication ("tautness"), must be considered. In a condition that is relatively rare, surgeons should be cautious to make sure that the endoscopic procedure they perform is as safe and effective as the open approach.

FURTHER READING

Becmeur F, Talon I, Schaarschmidt K, et al. Thoracoscopic diaphragmatic eventration repair in children: About 10 cases. *J Pediatr Surg* 2005; 40: 1712–15.

Cherian A, Stewart RJ. Thoracoscopic repair of diaphragmatic eventration. *Pediatr Surg Int* 2004; 20: 872–4.

Flageole H. Central hypoventilation and diaphragmatic eventration: Diagnosis and management. *Semin Pediatr Surg* 2003; 12: 38–45.

Groth SS, Andrade RS. Diaphragmatic eventration (review). *Thorac Surg Clin* 2009; 19: 511–19.

Hu J, Wu Y, Wang J, et al. Thoracoscopic and laparoscopic plication of the hemidiaphragm is effective in the management of diaphragmatic eventration. *Pediatr Surg Int* 2014; 30: 19–24.

Wu S, Zang N, Zhu J, et al. Congenital diaphragmatic eventration in children: 12 years' experience with 177 cases in a single institution. *J Pediatr Surg* 2015; 50: 1088–92.

Yazici M, Karaca I, Arikan A, et al. Congenital eventration of the diaphragm in children: 25 years' experience in three pediatric surgery centers. *Eur J Pediatr Surg* 2003; 13: 298–301.

16

Principles of lung surgery

SALEEM ISLAM, JAMES D. GEIGER, STEVEN S. ROTHENBERG, AND SHAUN M. KUNISAKI

PRINCIPLES AND JUSTIFICATION

Lung surgery is a relatively rare event in childhood, since acquired lesions such as carcinoma or chronic infections are much less common than in adults. Many operations are performed for congenital problems, and less commonly for infectious etiologies. **Table 16.1** lists the indications for pulmonary resection in children.

Table 16.1 **Pediatric lung conditions requiring surgery**

Congenital	Acquired
Sequestration	Acquired lobar emphysema
Cystic adenomatoid malformation	Bronchiectasis
Lobar overexpansion	Apical (or other) blebs
Lung cysts (single, multiple)	Metastatic lesions
Hamartomas	Fungal infections (lobar)
Congenital pulmonary insufficiency (needing biopsy for diagnosis)	Diffuse pulmonary disease

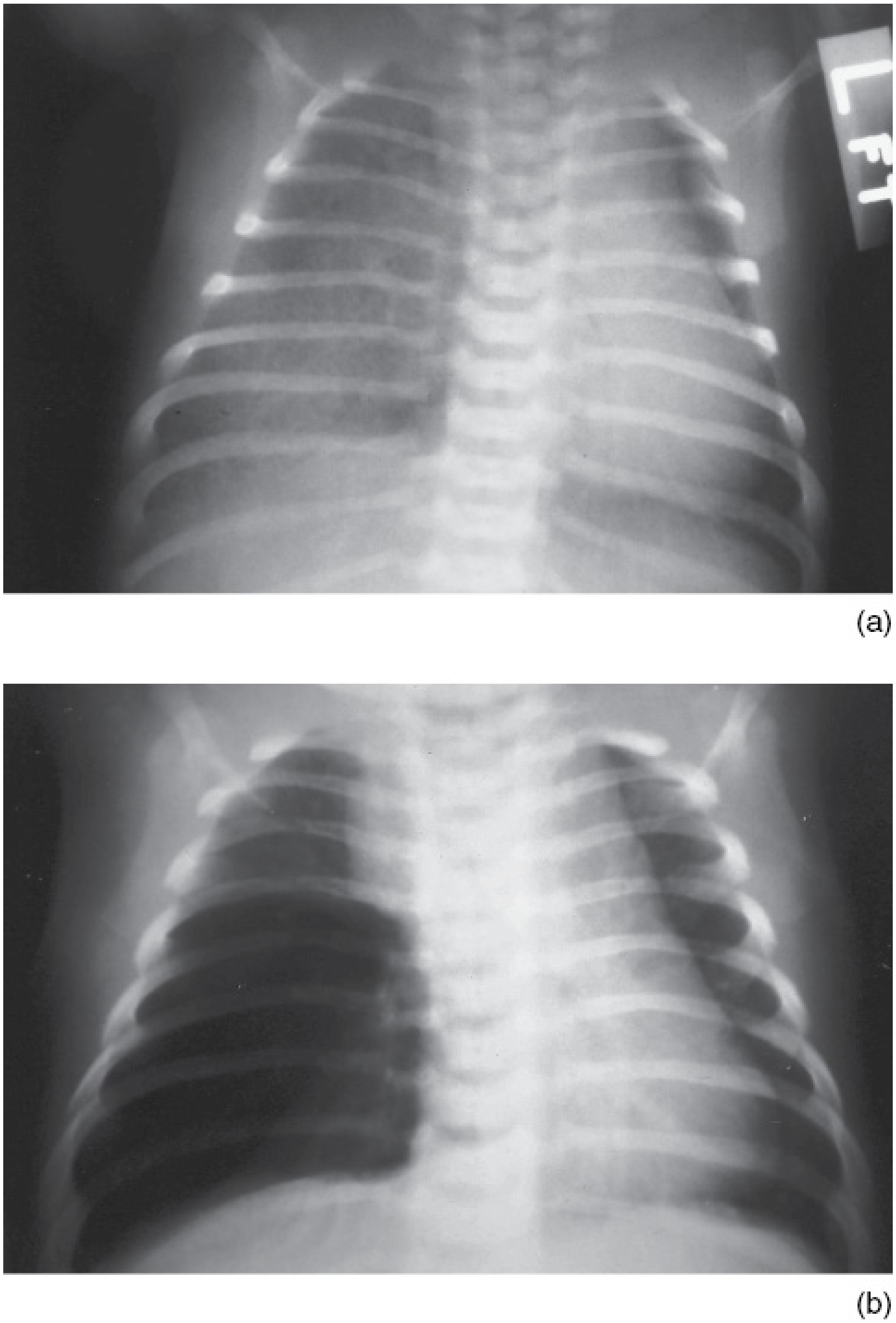
In general, the principles of lung surgery are similar in adults and children. Children usually have a greater physiologic reserve and withstand resections better than older patients. In the first seven years of life, there is ongoing alveolar development, which may diminish the impact of resection. Depending on the indications and the anatomic location for pulmonary resection, the approach and operative principles have to be adjusted. Minimally invasive techniques are being used more frequently in children and there is growing experience in thoracoscopic pulmonary resections in this age group. However, in cases of severely altered anatomy, such as inflammatory conditions, an open procedure is usually performed.

Complications of lung surgery in children are typically less than in adults. Major bronchial stump air leaks are rare in healthy pediatric patients. Scoliosis and/or the development of chest wall deformities are potential long-term complications unique to children who have undergone open thoracotomy and a minimally invasive approach can avoid this complication. Children with inflammatory conditions such as lung abscess, pneumonia, cystic fibrosis, and bronchiectasis are at greater risk for postoperative complications such as prolonged air leak.

We will first discuss the major conditions that result in the need for pulmonary resections in infants and children. These mainly consist of congenital problems, such as lobar overexpansion, pulmonary airway malformations, and sequestrations.

CONGENITAL LOBAR OVEREXPANSION (EMPHYSEMA)

Lobar overexpansion can be found in neonates and infants. This may be acquired from mucous plugs or structural abnormalities obstructing a portion of the airway. Congenital lobar overexpansion is caused by absence of the bronchial cartilage in 35% of cases, leading to ball valve-type air trapping. Occasionally, extrinsic compression (e.g. congenital heart anomalies) can lead to distal air trapping and overexpansion. Bronchogenic cysts are also seen in association with this condition. A number of cases result from alveolar hyperplasia in a specific lobe of the lung leading to an emphysematous appearance. The symptoms arise from compression of other thoracic and mediastinal structures, which may occur acutely or chronically. Some infants do not require surgical intervention and remain stable without removing the lesion. Of those who need an operation, 50% will develop symptoms within a few days of birth, while the remaining present a few months later. Chest radiograph will show a hyperlucent area in the affected chest with a variable degree of compression of the mediastinal structures. The diaphragm is flattened on the affected side (**Figure 16.1a and b**). Involvement is usually restricted to the upper lobes (42% left upper, 21% right upper, 35% right middle), with less than 1% involving the lower lobes. Differential diagnosis includes pneumothorax, pulmonary airway malformation, pneumatocele, and atelectasis.



16.1a,b

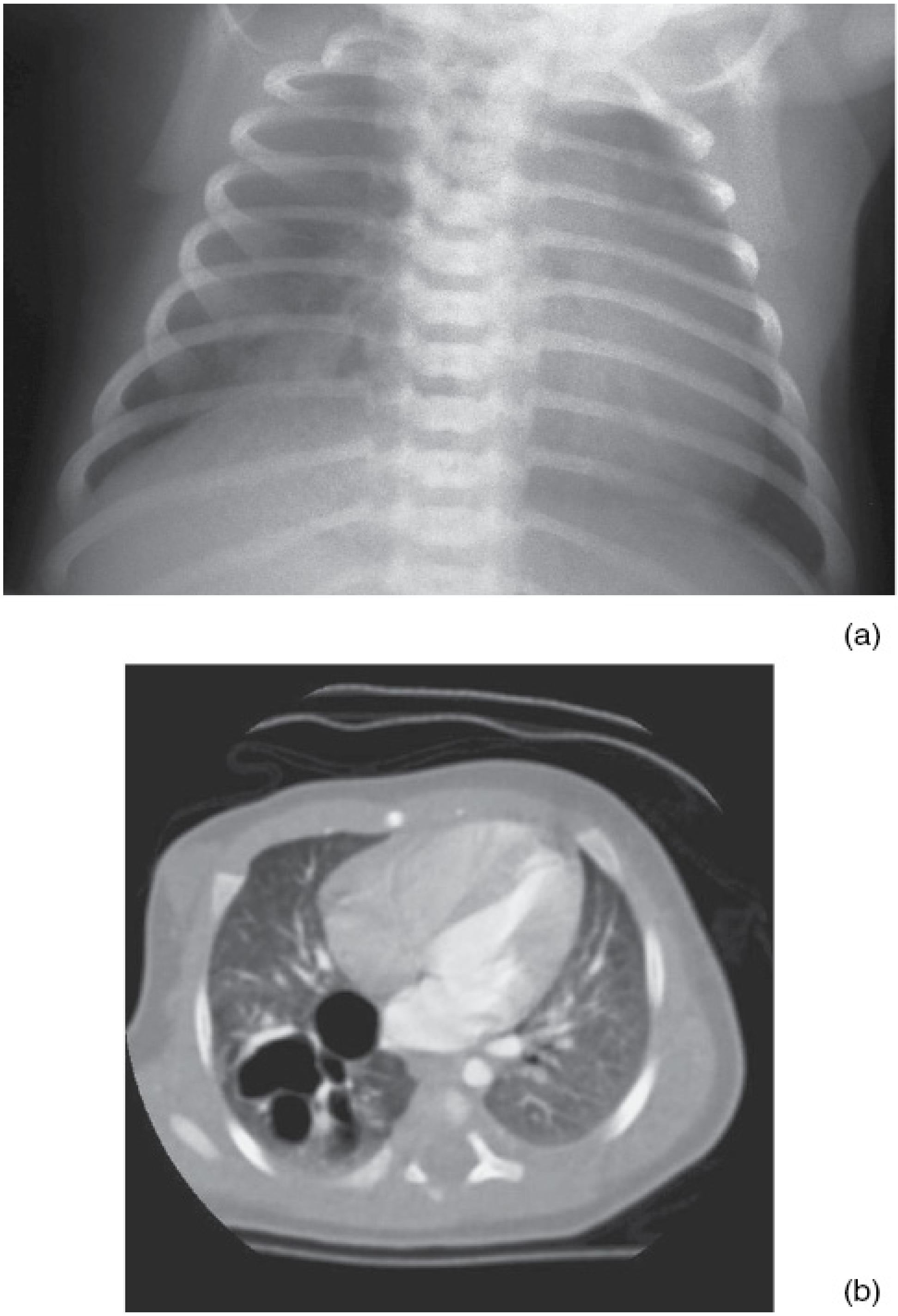
Additional studies are usually unnecessary, but ventilation perfusion scans are occasionally used and show delayed uptake and poor vascular supply to the affected lobe. In some instances, it is necessary to perform an emergency thoracotomy for life-threatening compression. The lobe will usually herniate out as soon as the chest is opened, with immediate clinical improvement. Importantly, the anesthesia team should not overventilate the patient, but should use rapid small tidal volumes; and the surgeon must be prepared to rapidly open the chest, as deterioration may occur with positive pressure ventilation.

Resection usually involves a lobectomy. If possible, bronchoscopy should be performed before resection to rule out a mucous plug or another obstructing lesion.

CONGENITAL PULMONARY AIRWAY MALFORMATIONS

Congenital pulmonary airway malformations (CPAMs, previously known as congenital cystic adenomatoid malformations (CCAMs)) are lesions that are mostly diagnosed with prenatal ultrasound. They form 25% of all congenital lung malformations but are still rare. Some become large enough that they cause lung hypoplasia and can also impede caval blood return, thus causing polyhydramnios and hydrops fetalis. Some of the affected fetuses will be stillborn. In selected centers, fetal thoracotomy and lung resection are performed in those who develop hydrops, with resultant improved survival. CPAMs are found in the left lower lobe in 25%, left upper lobe in 20%, right lower lobe in 19%, and right upper lobe in 10%.

Some neonates have respiratory distress at birth, and they can present with severe pulmonary hypertension requiring extracorporeal membrane oxygenation (ECMO) for stabilization prior to resection (extremely rare). Chest X-ray will often reveal an irregular cystic mass in the affected lung, which may involve the entire chest and have mediastinal shift (**Figure 16.2a**). In most cases, however, the infant is asymptomatic at birth and the postnatal chest X-ray is relatively normal. In these cases, a subsequent chest computed tomography (CT) can be obtained after discharge (generally performed at 1–2 months of age) and used to confirm the prenatal diagnosis (**Figure 16.2b**). In patients who are in no distress after birth, resection is usually delayed until the child is between 3 and 18 months of age, although most series now show advantages to resection prior to 1 year of age. Operation at a younger age may be easier due to the lack of inflammation. The risks of infection and cancer remain the major indications for elective resection. Bronchoalveolar carcinoma, pleuropulmonary blastoma, and rhabdomyosarcoma have been reported to arise in these lesions, but the incidence is considered very low.

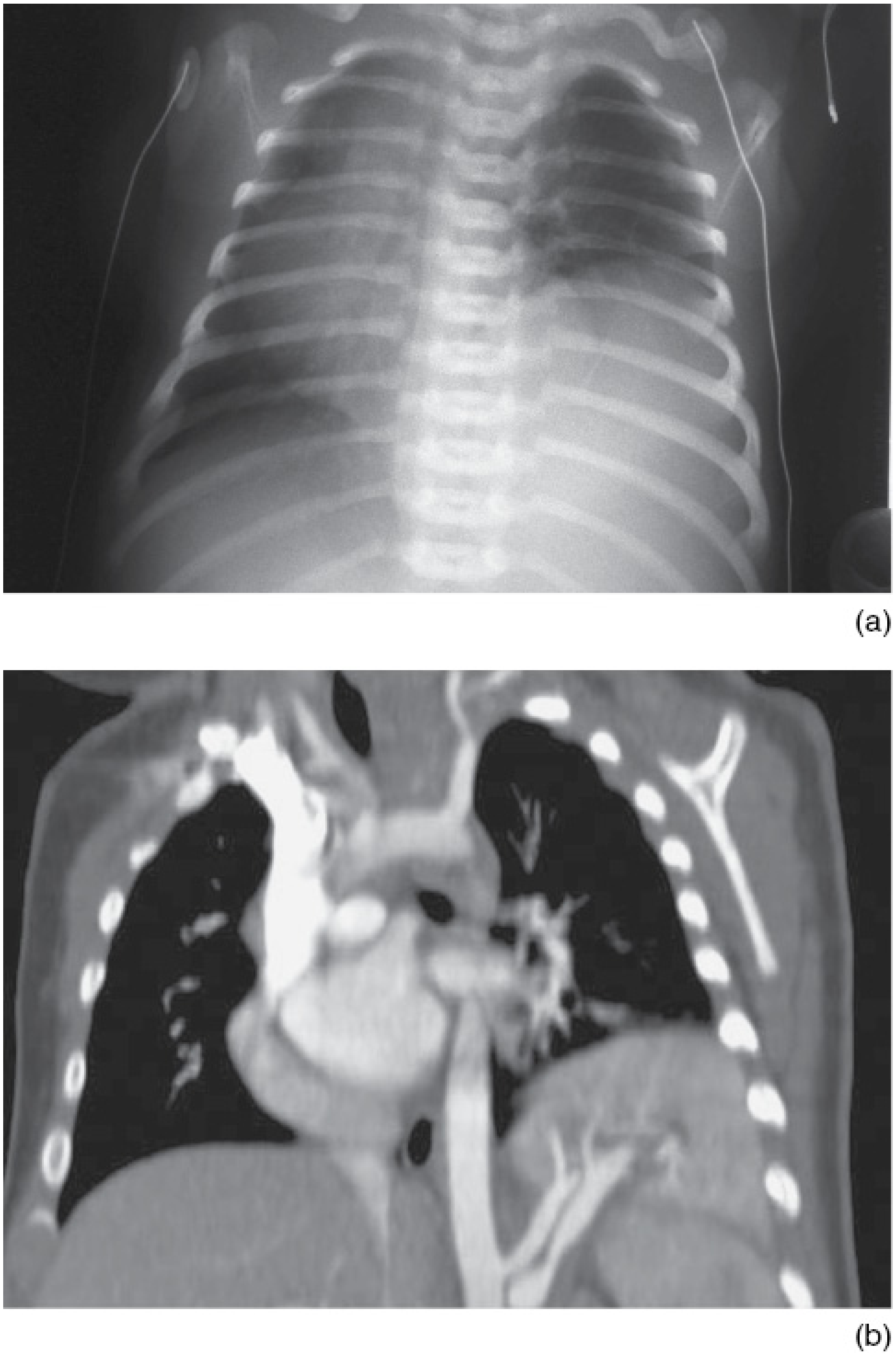


16.2a,b

In symptomatic patients, resection is performed in the early neonatal period. The most common procedure is a lobectomy, although segmentectomy has also been reported. It is considered vital to resect the entire CPAM due to recurrence or prolonged air leak. In some cases, the CPAM may involve multiple lobes, and in very rare cases pneumonectomy may be considered, although most would perform segmental resections in order to spare lung tissue.

SEQUESTRATION

A bronchopulmonary sequestration is a congenital malformation in which a portion of the lung receives systemic arterial supply and has no bronchial communication. There are two types, intralobar and extralobar sequestrations. Most large series describe a preponderance of extralobar lesions (**Figure 16.3a and b**). These may coexist with other malformations such as bronchogenic cysts, CPAM, and congenital heart lesions. Extralobar sequestrations are also associated with diaphragmatic hernias and eventrations. They may also occur in the abdomen, usually in the vicinity of the left adrenal gland. Most intralobar sequestrations are found in the lower lobes, with only 15% in the upper lobes. Presentation of intralobar lesions may be in the form of recurrent pneumonias, although increasingly they are being diagnosed by prenatal ultrasound. Extralobar sequestrations are usually asymptomatic and can be diagnosed on prenatal ultrasound, incidentally on a routine chest X-ray, or during the repair of a congenital diaphragmatic hernia or eventration. They also may be found incidentally as a posterior mediastinal mass. It is important to remember that there may be a communication of the sequestration with the foregut (esophagus or stomach). During resection, these communications should be sought and repaired if present. The vascular supply arises from the abdominal aorta in 85% and needs to be carefully ligated at the outset. It is usually found in the inferior pulmonary ligament, and care must be taken when mobilizing the structure. There is no bronchial attachment, which makes the resection easier. Intralobar sequestrations usually have a clear demarcation from the normal lung parenchyma and this forms a good plane of dissection. Lobectomy for intralobar lesions is the procedure of choice.



16.3a,b

PREOPERATIVE ASSESSMENT AND PREPARATION

If the patient meets the indications for lung surgery, pulmonary function should be optimized and infections should be controlled as much as possible with preoperative antibiotics, especially in the patient with cystic fibrosis (CF). Lobectomy and non-anatomic resections less than a lobectomy are well tolerated in healthy children. In patients with CF or other generalized pulmonary diseases in which lung reserve is reduced, resections, especially lobectomy, may not be tolerated. Preoperative workup in older children may include measurement of forced expiratory volume in the first second (FEV1) and forced vital capacity (FVC) to assess potential effects of resection, although this is not done in most otherwise healthy children. Most patients should have a minimum baseline measurement of hemoglobin, and appropriately cross-matched blood should be available in more complex resections (see Chapter 4 for additional discussion).

Anesthesia

Many lung operations in children can be successfully performed using a standard endotracheal tube without single lung ventilation. If required, single lung ventilation can be accomplished in infants and young children by either selective mainstem intubation or the use of bronchial blockers, which usually make the mainstem intubation technically easier and more reliable. In the older child, the use of double-lumen tubes (when at least a 7 mm endotracheal tube can be utilized) will allow for selective ventilation. For many simple thoracoscopic procedures, gentle insufflation at pressures of 5–10 mmHg will be enough to collapse the lung sufficiently, and selective ventilation may not be needed. However, for lobectomies, it is preferable to have the ipsilateral lung decompressed.

Postoperative pain relief after a thoracotomy is another important consideration, and the use of thoracic epidurals should be widely considered. Epidurals facilitate anesthetic delivery intraoperatively and decrease postoperative narcotic requirements. Intercostal nerve blocks can also be performed or, alternatively, a pleural catheter can be placed for delivery of local anesthetic.

Monitoring of children undergoing major lung surgery intraoperatively may involve the placement of an arterial line and a central venous line, but these are dependent on the patient's preoperative condition and on surgeon and anesthesiologist preference, and are not considered mandatory.

OPERATIONS

These are discussed in two broad categories: The general principles of pulmonary resection and specific approaches to various lobectomies, and the indications and operations needed for specific conditions.

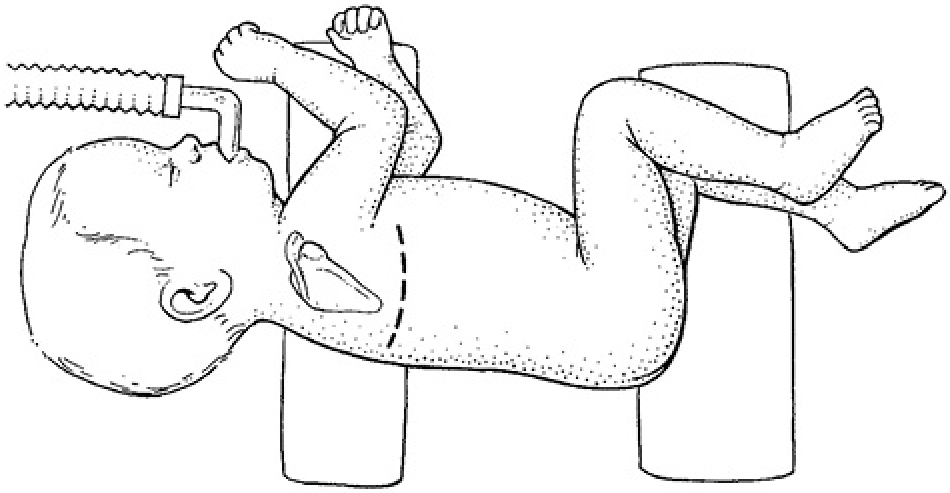
Principles of pulmonary resection

ACCESS TO THE LUNG

Operations on the lung can be performed in two ways, via thoracotomy or thoracoscopy. Thoracotomy can be performed by one of three approaches. The first is an anterolateral approach used mostly for open lung biopsies or wedge resections. The second is a posterolateral thoracotomy, which is the most common method used for open lung resections. A third approach through a prone position is employed to reduce spillover of infected secretions into the contralateral lung. However, with modern anesthesia techniques allowing single lung ventilation, as well as more effective antimicrobial therapy, this approach is less common. A median sternotomy is used by some surgeons when bilateral wedge resections are required, such as in osteosarcoma metastases.

The anterolateral thoracotomy is performed by elevating the patient with a roll 30–45° from the table. The incision is performed below the level of the nipple in the fourth, fifth, or sixth interspace, taking care not to injure the breast bud in a prepubertal female. The incision may be extended along the ribs toward the axilla as required. The pectoralis and intercostal muscles are then divided to enter the pleural cavity. After performing the desired resection or biopsy, a chest tube may be placed a couple of interspaces below the incision. The ribs are approximated loosely with an absorbable pericostal stitch (polygalactin). The muscle fascia is approximated with a running absorbable suture, followed by subcutaneous closure with either a running or interrupted suture and subcuticular closure of the skin (both with absorbable suture).

Posterolateral thoracotomy is performed with the patient in the lateral decubitus position. An axillary roll is used and appropriate padding of the legs is placed. The upper arm is allowed to lie on the same side with support to prevent excessive stretching of the arm as well as the brachial plexus. A wide preparation is done from the vertebral column posteriorly to the sternum anteriorly (**Figure 16.4**). The nipple and areola are marked, as is the tip of the scapula, to help guide the incision. A gently curved thoracotomy incision is performed in the interspace chosen. In most cases, a muscle-sparing approach can be employed (see Chapter 8 for details), in which the serratus anterior is retracted anteriorly or detached from the ribcage and the latissimus dorsi is reflected posteriorly. These muscles may be partially or completely divided as needed to gain wider access during the operation. Care is taken not to divide the paraspinal muscles, but to free them up longitudinally. This move, as well as avoiding division of the trapezius and rhomboids, may help in reducing the development of scoliosis. The ribs are held apart with a self-retaining metal retractor (Finochietto). After the resection is performed, a chest tube may be placed a couple of interspaces below the incision. Pericostal sutures are placed in an interrupted fashion and appropriately secured avoiding excessive approximation of the ribs. Fascia and skin are closed as described previously.

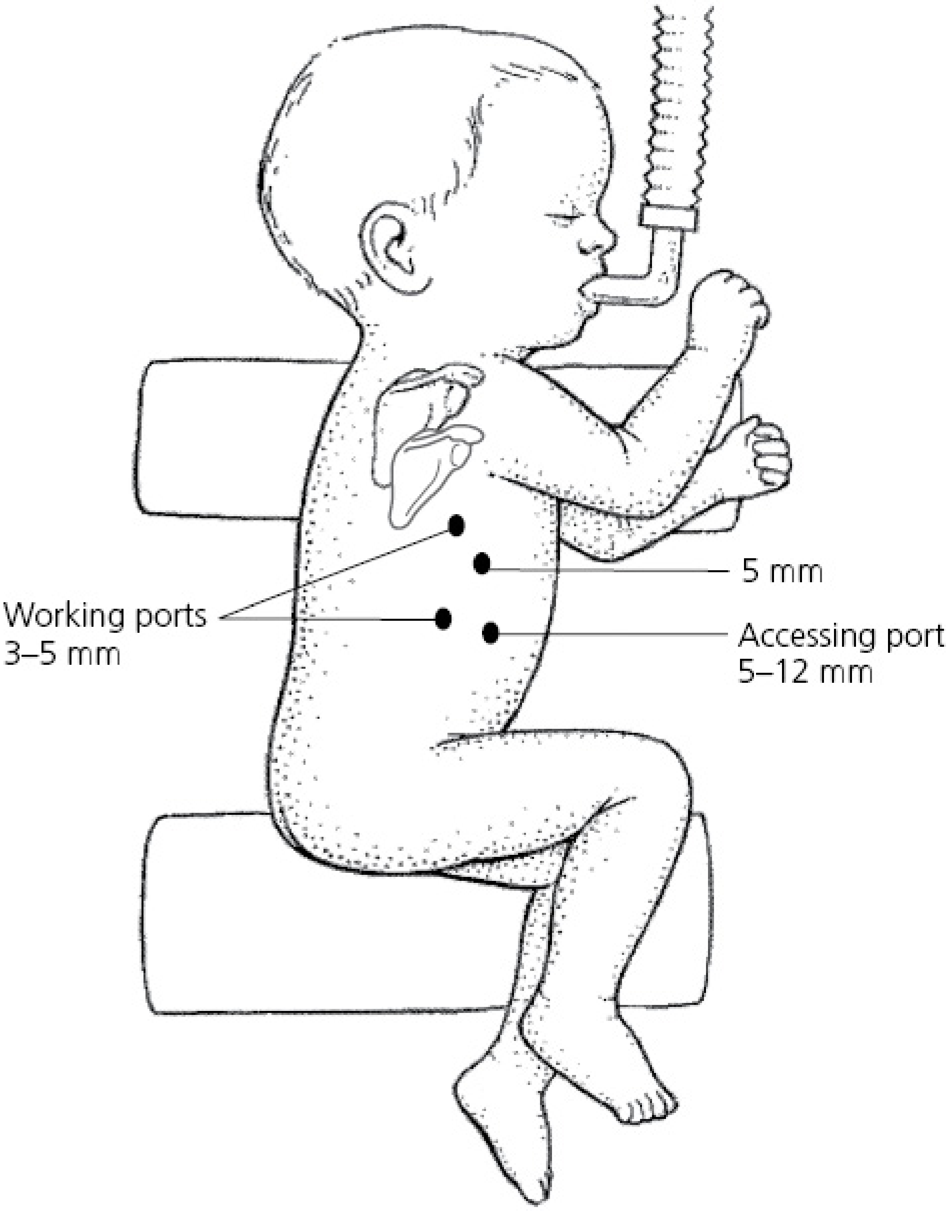


16.4

Thoracoscopic approaches are now being increasingly employed. Lung biopsies, wedge resections, and lobectomies can be performed using this approach. Improved optics with high-definition cameras and brighter light sources have made visualization excellent, and the development of safe vessel sealing devices for dissection and control of vessels up to 7 mm in diameter has allowed these resections to be performed safely. The clear benefits in cosmesis, decreased pain and length of stay, and less chest wall deformity and scoliosis have been the driving forces. For thoracoscopic resections of any kind, the patient is placed in a lateral decubitus position, as described previously. The table may be rotated to the right or left as needed for exposure. Some surgeons prefer to have the patient in the position described for the anterolateral thoracotomy. It is important to make full use of gravity as a retractor and the surgeon should vary the position accordingly. Single lung ventilation should be used to facilitate exposure for lobectomy, although CO_2 insufflation suffices for other procedures. The chest is prepared as for a thoracotomy and a 5 mm incision made in the mid-axillary line in the fourth to sixth interspace. A Veress needle is placed into the chest carefully just above a rib to avoid the neurovascular bundle.

Insufflation with CO_2 to a pressure of 3–7 mmHg at a flow of 1–3 L/min, at least initially, facilitates exposure and creates a functional working space.

A 4 mm or 5 mm port is placed and the 4 mm or 5 mm 30° telescope is introduced. The remaining port sites are then chosen based on the specific anatomy and which lobe is to be operated upon. Usually, two additional ports are placed to facilitate triangulation and dissection of the fissure and lobe (**Figure 16.5**). These positions are variable and can move one or two interspaces up or down as needed, and for lobectomy are typically located in the anterior axillary line. A fourth access site may be used for retraction if necessary. In larger patients (>10–15 kg) it may be desirable to use a 12 mm endoscopic stapler and thus require a 12 mm port. However, these devices are generally much too large for the chest cavity of patients under 10 kg and should be avoided to prevent inadvertent injury to surrounding structures. The surgeon should also avoid mass division of structures across the fissure with the stapler. Artery, bronchus, and vein should always be dissected out and taken separately. One of the port sites may need to be enlarged slightly to remove the specimen. Upon completion of the procedure, a chest tube may be placed through one of the dependent port sites and secured. The fascia may be closed with an absorbable stitch and the skin approximated by suture or other means.



16.5

LUNG BIOPSY AND WEDGE RESECTION

This procedure is performed for diagnostic or therapeutic purposes. Indications for biopsy include infectious processes, diffuse parenchymal diseases, and presence of lung nodules where a diagnosis of an inflammatory or malignant process is being considered. Those patients who have diffuse disease may not tolerate single lung ventilation, and in these cases the procedure will have to be performed with the lung expanded.

Thoracoscopic procedures allow visualization of the entire lung making a small wedge resection easy to perform with endoloops or a stapling device. An open operation with a small anterolateral thoracotomy is also a reasonable technique for a wedge biopsy with a stapling device. A persistent air leak after a non-anatomic resection can be controlled by a second firing of the stapling device after removing the knife blade, or by oversewing the suture line. Buttressed staplers or fibrin sealant may also decrease the risk of air leaks. Chest closure is performed in the same way as described previously.

Non-anatomic wedge resections may also be performed for metastatic disease of the lung. In general, an open thoracotomy is preferred for osteosarcoma metastatic disease, to allow palpation of the lung parenchyma and to remove as many lesions as possible, which is thought to improve survival. Usually both lungs are evaluated in this fashion for metastatectomy.

In other tumors, it may be reasonable to utilize a thoracoscopic approach. Smaller or deep parenchymal lesions may not be accessible via thoracoscopy. Some authors have described using CT-guided marking of the lesions with methylene blue ink, a blood patch, or a needle localization immediately preoperatively to facilitate localization and resection with thoracoscopy.

Apical bleb disease leading to recurrent or persistent pneumothorax can also be treated thoracoscopically. The apex is visualized and wedges of parenchyma with blebs are removed using an endoscopic stapling device. In most cases, it is best to use these devices with the vascular load, which has smaller staples and thus results in less bleeding. Again, if there is an air leak from the staple line, a further firing of the device after removing the blade or using a stapler that has been reinforced with bovine pericardium may be helpful. Fibrin glue can be applied over the staple line as an adjuvant to help control air leaks.

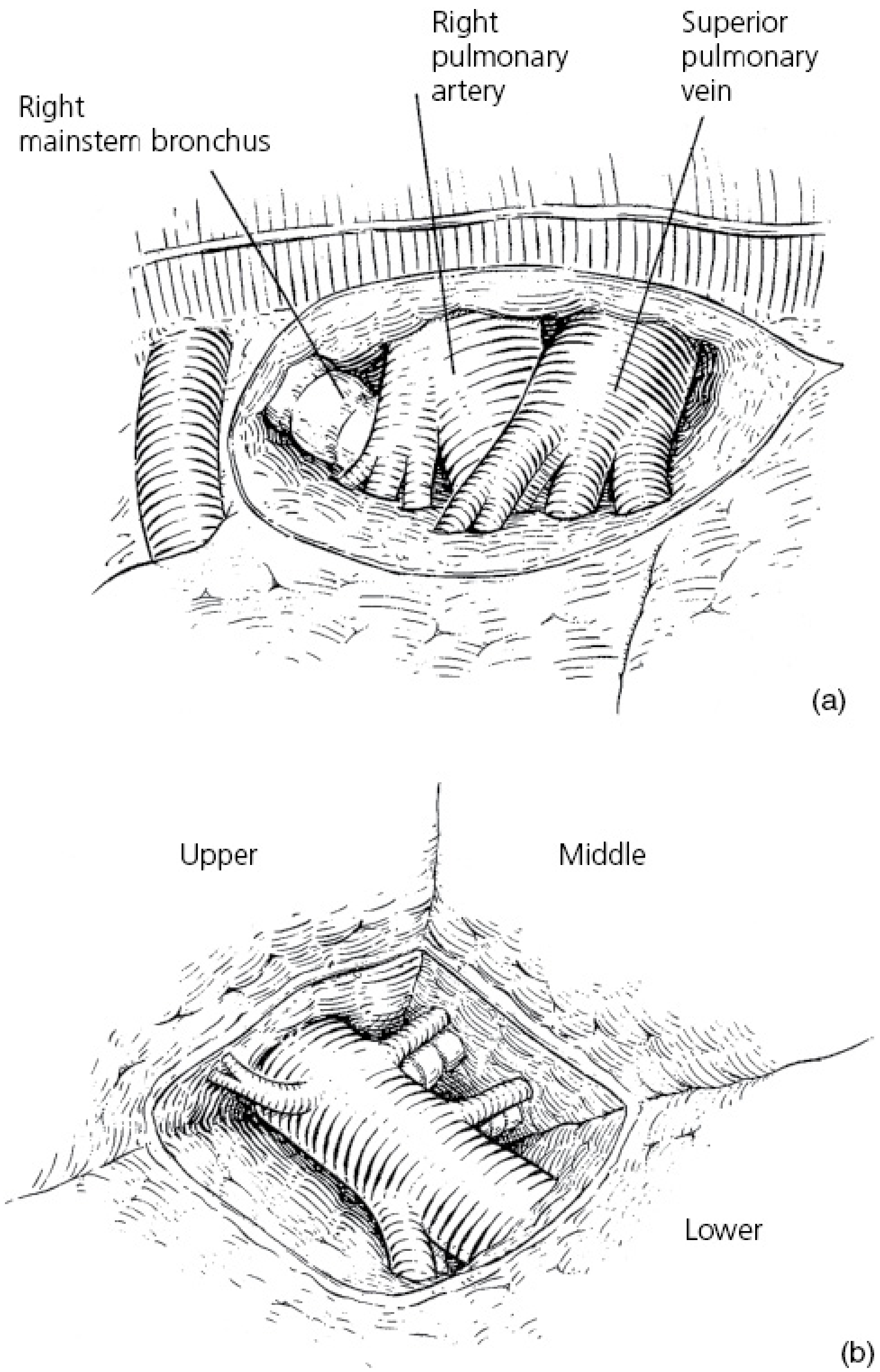
LOBECTOMY

The principles of lobectomy are similar in children and adults. The most important considerations are adequate visualization and exposure of the hilar structures, i.e. the blood vessels and bronchus. In most instances, it is better to dissect out and control the pulmonary arterial branches first and then, depending on the lobe and anatomy, the venous drainage and the bronchus. Occasionally, as in the case of a severe, purulent infectious process, it is better to divide the bronchus first.

The anatomic considerations involved in the removal of different lobes will now be discussed in detail. The surgeon must have a complete understanding not only of the normal anatomy but also of the variations in anatomy that are frequently encountered during lung resections.

Right upper lobectomy

The patient is positioned for either a posterolateral thoracotomy via the fifth interspace or a thoracoscopic approach. The lung is retracted posteriorly and the pleura covering the hilum of the right lung is opened anteriorly and posteriorly to a level below the right mainstem bronchus. Care is taken to avoid injury to the right phrenic nerve. It should be noted that considerable variation in vascular anatomy occurs, but the usual pattern is shown in **Figure 16.6a**. The first vessel to be encountered will be the superior pulmonary vein which will have the upper and middle lobar branches. The main pulmonary artery is identified behind the superior pulmonary vein, and dissection is carried out peripherally to expose the superior and inferior pulmonary arterial trunks (**Figure 16.6b**). The superior pulmonary arterial trunk, with its branches to the anterior, posterior, and apical segments, is exposed and then branches individually ligated. The ligation of the vessels can be aided by dissecting into the lung parenchyma to gain added length. This technique also allows one to ligate arteries of smaller caliber more effectively using clips or energy devices. The superior lobe vein is identified and dissected laterally, usually exposing three segmental pulmonary veins. The middle lobe veins entering the superior pulmonary vein must be identified and preserved. It is often easier and safer to ligate the individual venous branches peripherally and in the lung parenchyma. The oblique fissure is then opened and completed between the upper and lower lobes. This permits the dissection of the posterior ascending segmental pulmonary arterial branch, which ordinarily comes off the pulmonary artery after the middle lobe branches. All the vessels are best managed by ligation with non-absorbable suture, as well as suture ligation proximally.


Right mainstem bronchus
Right pulmonary artery
Superior pulmonary vein
(a)
Upper
Middle
Lower
(b)

16.6a,b

After division of the arterial and venous branches of the right upper lobe, adventitial tissue surrounding the bronchus is cleared away. Stay sutures of non-absorbable material are placed on either side of the bronchus and it is divided about 1–2 cm from the mainstem in order to avoid a long stump, which may accumulate secretions. After division of the bronchus, the stump is closed with interrupted sutures of 3/0 or 4/0 monofilament or braided non-absorbable material.

THORASCOPIC APPROACH

When performing the operation with video-assisted thoracic surgery (VATS), a bipolar sealing device is probably the safest and fastest technique to ligate and divide the vessels. In general, it is safest to make two separate seals at least 3–4 mm apart and sharply cut the vessel between them. The initial cut should be part way through the vessel to ensure that the seals are secure prior to the vessel retracting and vascular control is lost. If there is bleeding, the seals can be readdressed. The 3 mm vessel sealer (JustRight Surgical; Louisville, CO) will safely seal vessels to 5 mm and the 5 mm sealer (LigaSur; Boulder, CO) up to 7 mm.

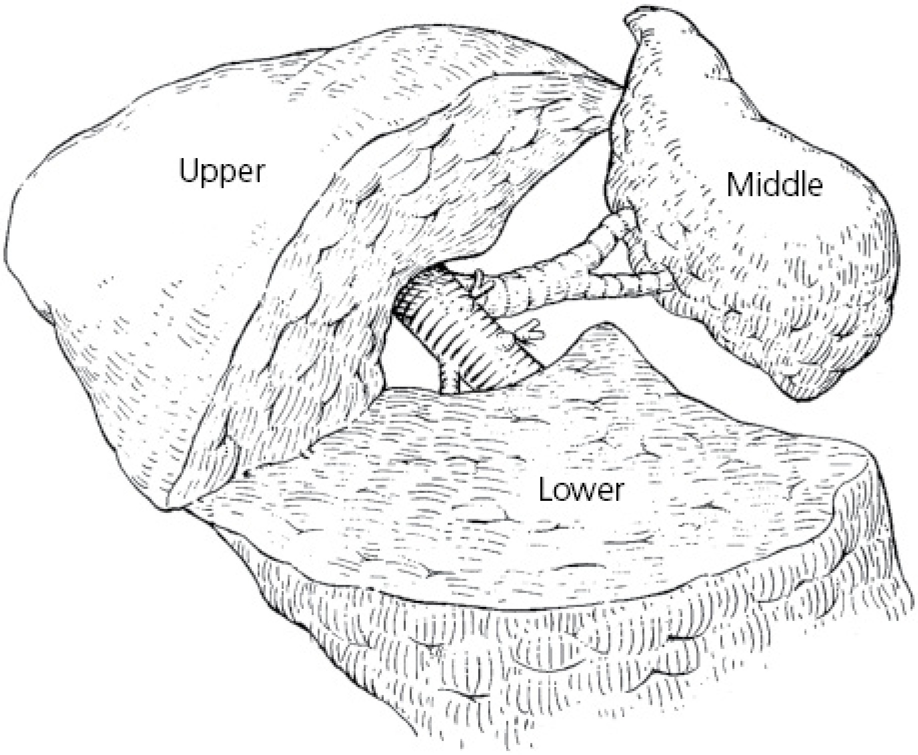
A right upper lobectomy is probably the most difficult to perform thoracoscopically because of the wide distribution of the segmental vessels and the need to preserve the middle lobe vessels and bronchi. Therefore, the surgeon must have a clear understanding of the anatomy and the techniques before attempting this thoracoscopically. The dissection begins similarly, with the exposure of the superior vein and branches; it is sometimes easier to ligate and divide these branches before the arterial branches. Teasing the lung off the vessels is a good technique to use in these cases to gain length and ensure no compromise to the main vessel. This maneuver also allows for thermoligation of smaller vessels which may be somewhat safer. Completion of the minor fissure can be performed with vessel sealing in most cases without air leak. In larger patients, an endoscopic stapler may quicken the procedure.

Thoracoscopically, there are a number of safe choices for dealing with the bronchus. This dissection is usually facilitated by the fact that in most cases the bronchial tree mimics the arterial tree. Therefore, having successfully ligated the arterial branches, the corresponding bronchus should now be exposed. Options for taking the bronchus include sharp division and closure with interrupted polydioxanone (PDS) or prolene sutures. The primary disadvantage of this is that it creates an open airway with loss of tidal volume which may make ventilating more difficult for anesthesia, especially when perfect single lung ventilation has not been achieved. This technique is also significantly more time consuming. In smaller infants 5 mm endoscopic clips (regular or Hem-O-Lock, Teleflex Medical, Reading, PA) can be used, primarily at the segmental level. There is now a 5 mm endoscopic stapler (JustRight Surgical, Louisville, CO) available which works well to take the mainstem bronchus in patients up to approximately 10 kg. In larger patients a 12 mm endoscopic stapler is probably the best choice. Torsion of the right middle lobe has been reported after an upper lobectomy, and the right middle lobe should be sutured to the right lower lobe if the major fissure is complete after a right upper lobectomy is performed.

After closure, warm saline is poured into the chest and the stump is tested for a leak by applying a pressure of 30–40 cmH_2O with the ventilator. Surrounding pleura may be used to reinforce the stump and possibly promote healing. A chest tube is placed and secured as described previously.

Right middle lobectomy

The arteries to the right middle lobe are best exposed through the oblique (minor) fissure between the upper and middle lobes (Figure 16.7). After development of the interlobar fissure, one or two middle lobe vessels are usually encountered near the posterior aspect of fissure near the confluence with the major fissure. These are mobilized and ligated as previously described by dissecting into the middle lobe parenchyma as necessary. The lung is retracted posteriorly to expose the anterior hilum, and one or two right middle lobe veins are found in the area mentioned previously, joining the superior pulmonary vein. Once the middle lobe has been separated from the upper lobe and both arterial and venous branches divided, the lobe is retracted anteriorly and the bronchus is dissected and divided. The stump is controlled in the fashion described previously. Care must be taken to avoid inadvertent injury to the ascending posterior segmental artery of the upper lobe during division of the bronchus.



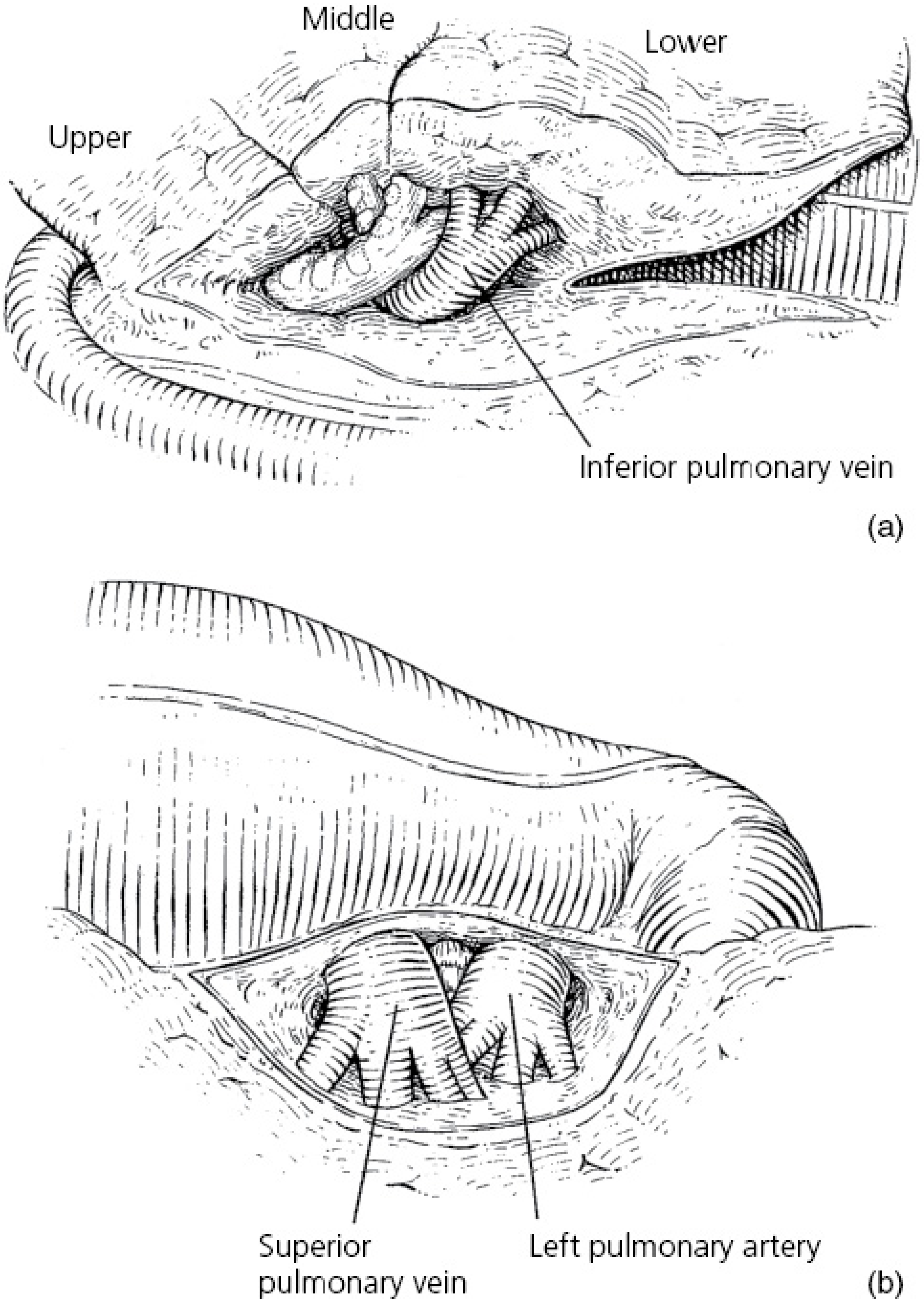
16.7

THORACOSCOPIC APPROACH

The working ports are shifted one or two interspaces up in the anterior axillary line for the middle lobectomy when done with VATS. The minor fissure is dissected and the arteries are encountered near the confluence of the major and minor fissures. The vein is located after posterior retraction of the lobe and dissection of the anterior pleura. The bronchus is identified near the apex of the lobe.

Right lower lobectomy

For performing a right lower lobectomy, a lower intercostal space may be used. The interlobar fissure is exposed by retraction of the upper and middle lobes superiorly and the lower lobe inferiorly. The branches of the interlobar portion of the right pulmonary artery are exposed and carefully identified. Just beyond the middle lobe arteries and opposite them, one or two superior segmental arteries supplying the superior segment of the lower lobe are encountered. These are divided after ligation. There is a remaining vessel to the basilar segments, which should be identified and similarly ligated. The lobe is then retracted anteriorly to expose the posterior hilum. The inferior pulmonary vein is exposed by opening the inferior pulmonary ligament and carrying the pleural dissection upwards to isolate and facilitate ligation of the inferior pulmonary vein (**Figure 16.8a and b**). After this, the right lower lobe bronchus can be easily identified by posterior retraction. Care is taken to keep the bronchial stump length short.


Middle
Lower
Upper
Inferior pulmonary vein
(a)
Superior pulmonary vein
Left pulmonary artery
(b)

16.8a,b

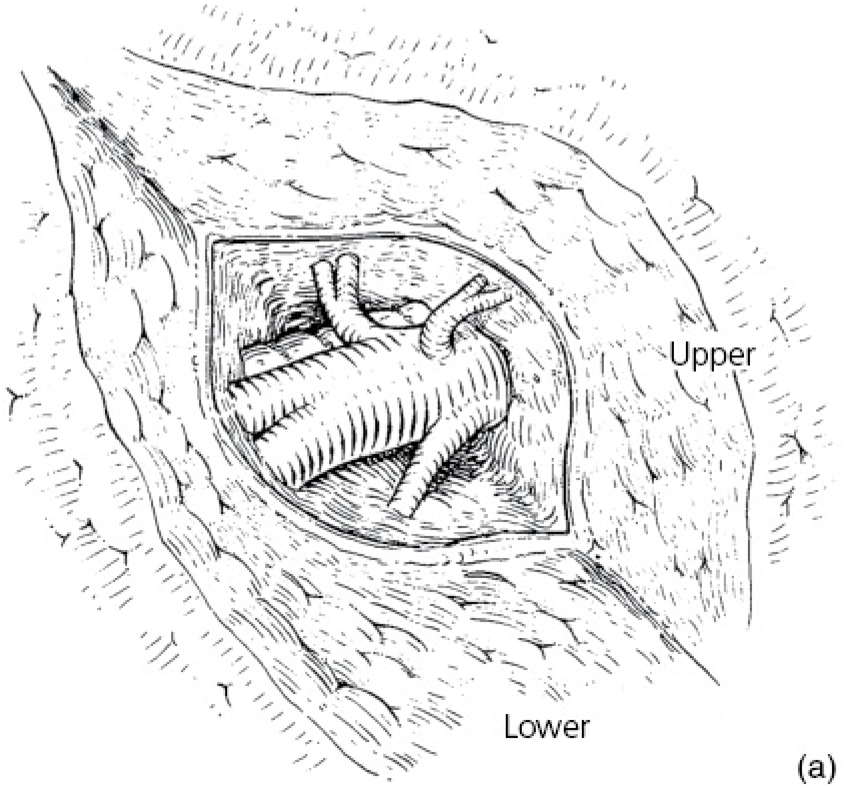
THORACOSCOPIC APPROACH

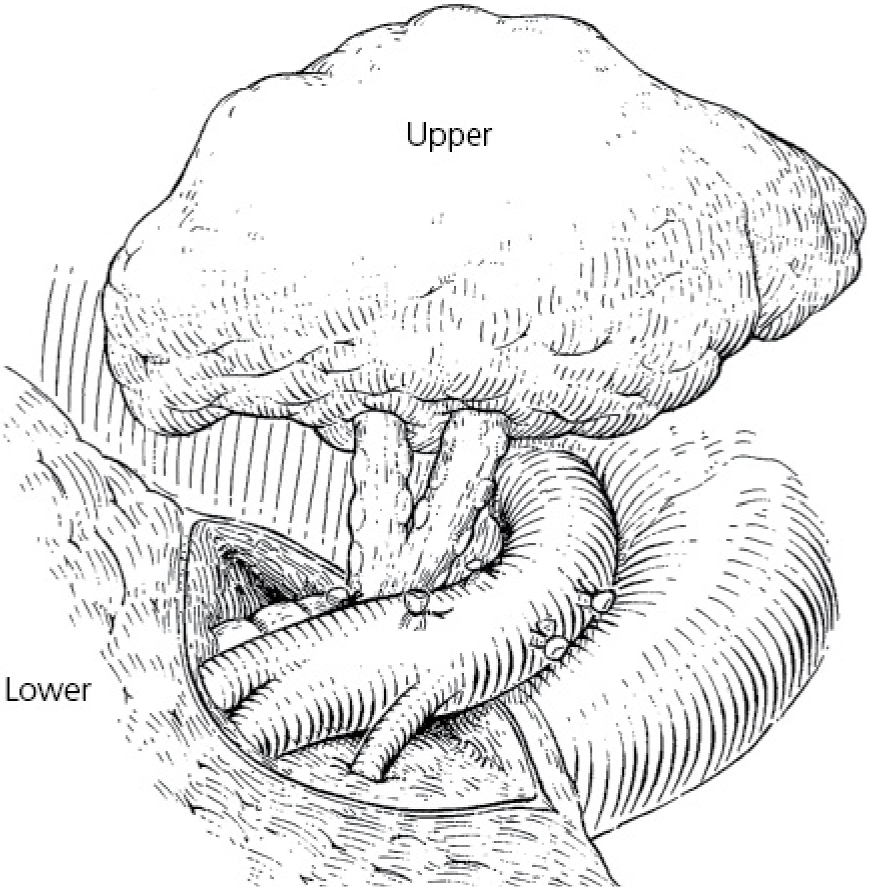
When performing this resection using the thoracoscope, the first move is typically dissection of the inferior pulmonary ligament to expose the inferior pulmonary vein. During this dissection, it is prudent to look for an aberrant systemic vessel as occurs in a sequestration. The vein is not ligated until after the arterial dissection to avoid congestion of the lobe. The upper and middle lobes are then retracted superiorly to expose the fissure, which is completed. This exposes the pulmonary artery to the lower lobe. Depending on the anatomy, the vessels may be taken at the segmental level or in some cases the main trunk to the lower lobe maybe addressed directly. Often it is helpful to take the superior segmental vessel first creating more length along the basal trunk. This trunk can be taken with the endoscopic stapler (5 mm or 12 mm), saving time by eliminating the need to dissect each branch separately. Otherwise the basal segmental vessels are individually dissected out, sealed, and divided as described previously.

Because division of the arterial branches exposes the bronchi, it is often easier to now address the bronchus. This can easily be dissected out, staying with the same exposure as for the artery. The surgeon just needs to be aware that the inferior pulmonary vein lies just behind the plane of dissection. The advantage of taking the bronchus first is that it eliminates the need to shift the lung and change exposure to visualize the vein. It is also much easier to address the inferior pulmonary vein once the bronchus is divided. As with the artery, it is often helpful to take the bronchus to the superior segment first to create greater length on the basal segments trunk. Depending on the size of the patient, the bronchus can be taken with clips or a 5 mm or 12 mm stapler. With the bronchus divided, the trunk of the inferior pulmonary vein is easily seen and managed. The lobe is held up and the vein can now be taken with staples, clips, or sealer. Generally, if the sealer is used, dissection must be carried toward the parenchyma to address the vein at its branching point so the vessels are of smaller caliber. If the stapler is used, care must be taken to ensure the stapler is 1–2 cm away from the base of the trunk so that proximal control of the vein is possible if there is a misfire.

Left upper lobectomy

After positioning the patient in the appropriate right lateral decubitus position, a posterolateral thoracotomy is performed. As for the right lung, the pleura overlying the hilum of the lung anteriorly is incised and carried superiorly and posteriorly below the level of the left mainstem bronchus. The left pulmonary artery is best identified anteriorly first, and then found as it courses superiorly and posteriorly to the upper lobe bronchus. Four to six branches of the left pulmonary artery to the upper lobe can be noted (**Figure 16.9a**). Anteriorly, the anterior, apical, and posterior segmental arteries are seen. The apical segmental artery may be encountered superiorly, and anterior segmental and lingular segmental branches are usually seen in the interlobar fissure (**Figure 16.9b**). After ligation of all these branches, the lung is retracted posteriorly and the left superior pulmonary vein is ligated just before it divides. Occasionally, the left superior and inferior pulmonary veins form a common vein, so before ligating the superior vein on the left side, the inferior vein should be identified separately. With anterior retraction, it is possible to see the bronchus to the upper lobe and lingula and divide these close to the origin as described.


Upper
Lower
(a)


Upper
Lower

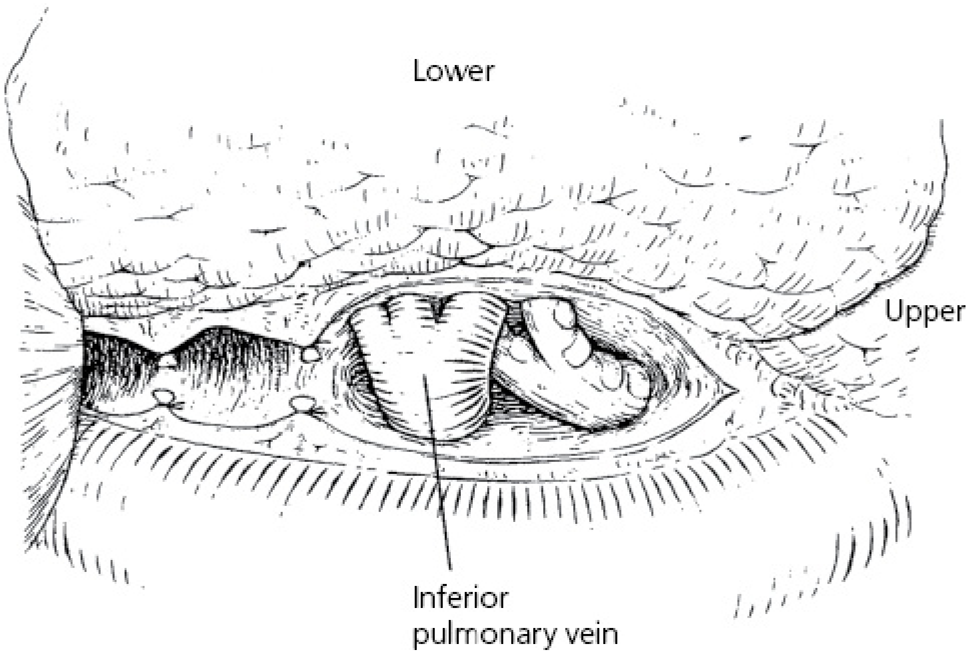
16.9a,b

THORACOSCOPIC APPROACH

The thoracoscopic approach is similar to that mentioned for the right upper lobe, with exposure of the pulmonary vein and then the arterial branches, as discussed in the open technique.

Left lower lobectomy

After gaining access to the chest, the interlobar fissure is exposed to identify the arteries. One or two arteries supply the superior segment, but care must be taken as the superior segmental artery may arise proximal to the lingular vessels. Thus, the lingular arteries must be identified in the course of this dissection. Following ligation of these vessels, the basilar portion of the left pulmonary artery may be divided just distal to the lingular arteries. After this, the lung is retracted anteriorly to expose the posterior hilum. The inferior pulmonary ligament is divided to expose the inferior pulmonary vein (**Figure 16.10**). The vein is then divided as described, taking care to be sure the superior lobe has good venous drainage. After vascular division, the lobe is retracted posteriorly and the bronchus isolated and divided as described previously.



16.10

THORACOSCOPIC APPROACH

Thoracoscopic left lower lobectomy is very similar to the right side, as described previously. The vein is isolated first by taking down the inferior pulmonary ligament; however, the vein is not ligated until after pulmonary artery ligation. The pulmonary arterial branches are then dissected in the fissure and ligated using a vessel sealing device, taking care to identify the lingular vessels and preserve them. In general the bronchus is then taken as in the right lower lobectomy and then the vein branches isolated are ligated at this point.

POSTOPERATIVE CARE

Important principles of postoperative care after lung surgery involve ensuring adequate pulmonary toilet. Pain control is critical to optimizing postoperative pulmonary lung function and may be provided by a thoracic epidural. The epidural, if used, is usually maintained for 48–72 hours postoperatively. A Foley catheter for bladder drainage may be needed with epidural pain relief. Pain may not be a prominent feature following thoracoscopic procedures, but often the chest tube is a major cause of postoperative pain and should be removed as soon as possible. The chest tube is maintained for 1–3 days, initially on suction and then on underwater seal. The tube is removed once there is no air leak and the drainage is minimal.

Antibiotics may be given for 24 hours. Some surgeons continue antibiotics until the chest tube is removed, although there are no good data to support this practice. Narcotics should be used judiciously and non-narcotic analgesia incorporated into the algorithms for pain control. Feeding is usually resumed the day after surgery.

Intensive care is required for patients with significant underlying lung disease and reduced pulmonary function. Most patients are able to go home within 3–4 days, although older children may need to stay longer because of pain issues. After thoracoscopic procedures, the hospital stays are usually shorter.

OUTCOME

Mortality following lung resection, including lobectomy or bilobectomy, should be minimal with modern surgical techniques and postoperative care. The only group at increased risk is those with significant other comorbidities, such as congenital heart disease, or those with additional pulmonary disease.

Mortality and morbidity after lung biopsy are mainly dependent on the underlying disease process. The risk of prolonged air leak with diffuse parenchymal disease also exists and can add significantly to morbidity.

Infectious complications are not common following lung resection for congenital lobar overexpansion, uncomplicated cystic adenomatoid malformation (CAM), or sequestrations. When there is a pre-existing infection or abscess, the risk is higher. Bronchial stump leaks are very rare compared to adults.

VIDEO

OPS_8e_video_16.1 Thoracoscopic left lower lobectomy for CCAM

FURTHER READING

Aziz D, Langer JC, Tuuha SE, et al. Perinatally diagnosed asymptomatic congenital cystic adenomatoid malformation: To resect or not? *J Pediatr Surg* 2004; 39: 329–34.

Azizkhan RG, Crombleholme TM. Congenital cystic lung disease: Contemporary antenatal and postnatal management. *Pediatr Surg Int* 2008; 24: 643–57.

Puligandla PS, Laberge JM. Infections and diseases of the lungs, pleura and mediastinum. In: Grosfeld JL, O'Neill JA, Coran A, Fonkalsrud E. (eds). *Pediatric Surgery*, 6th ed. Boston, MA: Mosby; 2006: pp. 1001–37.

Rothenberg SS. First decade's experience with thoracoscopic lobectomy in infants and children. *J Pediatr Surg* 2008; 43: 40–4.

Rothenberg SS. Thoracoscopic lobectomy. In: Holcomb GW, Georgeson KE, and Rothenberg SS (eds). *Atlas of Pediatric Laparoscopy and Thoracoscopy*. Philadelphia, PA: Saunders Elsevier; 2008: pp. 253–60.

Su WT, Chewning J, Abramson S, et al. Surgical management and outcome of osteosarcoma patients with unilateral pulmonary metastases. *J Pediatr Surg* 2004; 39: 418–23.

17

Empyema

JOSEPH SUJKA AND SHAWN D. ST PETER

HISTORY

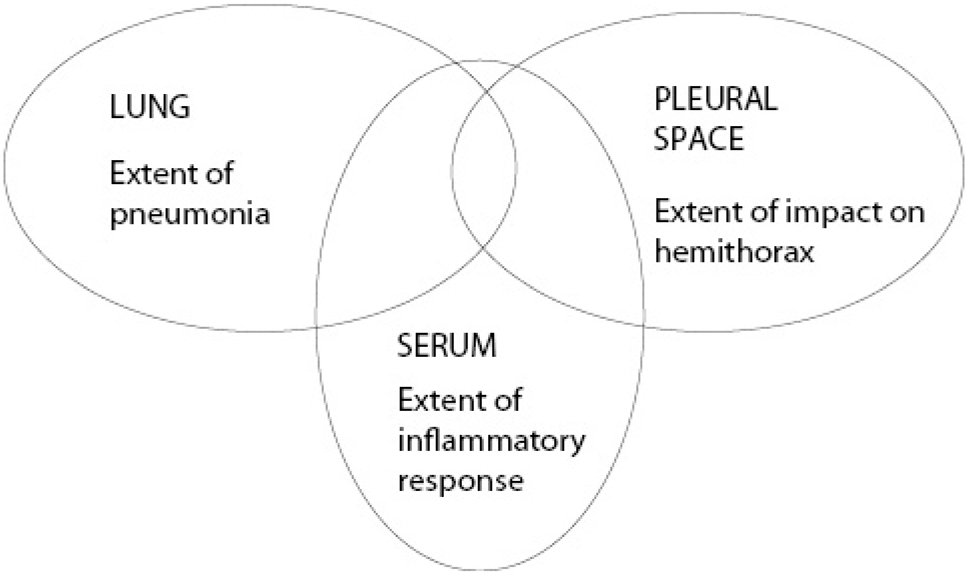
Empyema is defined as the accumulation of pus in a body cavity, derived from the Greek word *empyein* which means to "put pus in." In routine medical terminology it refers to pus in the pleural space. Usually a complication of inflammation or an infection within or adjacent to the pleural space, empyema rarely resolves spontaneously because host defenses are limited by the anatomy and physiology of the pleural space. In children the most common etiology is reaction to adjacent pneumonia; however, other sources include reaction to subphrenic abscess as well as extension of mediastinal, retropharyngeal, or paravertebral infectious processes. Empyema can also develop secondary to infection after thoracic surgery or trauma.

PRINCIPLES AND JUSTIFICATION

Overall rates of bacterial pneumonia have been declining in children but the incidence of complications, such as parapneumonic effusions (PPEs) and empyema, has increased. In the United States, pediatric pneumonia occurs at an estimated rate of 30–40 per 100 000. PPEs or empyema complicate pediatric pneumonia in 28–53% of cases. While pediatric empyema is less serious than it is in adults, where mortality can approach 20%, it still poses a considerable burden on hospitals and families. The natural progression of empyema is outlined with three or four stages of increasing complexity.

1. The *pre-collection stage* is characterized by a localized pleuritis.
2. In the *exudative stage* there is a simple effusion with clear, free-flowing pleural fluid with a low pleural white blood cell count.
3. The *fibrinopurulent stage*, or classic empyema, produces a complicated parapneumonic effusion. The infected pleural space results in a procoagulant environment leading to decreased fibrinolytic activity with a concomitant increase in fibrin deposition. Fibrin acts as the tangible matrix for septations and loculations. This is heralded by an increase in the white blood cell count of the fluid.
4. The *organization stage*, the most advanced phase, results in a thickened rind, which may entrap the lung, manifesting as chronic restrictive lung disease.

Despite being sequentially outlined, there is no certainty that each stage will progress to the next. More importantly, the stage of pleural disease may not relate to the degree of physiologic illness. The severity of illness is determined by the extent of underlying parenchymal disease and the extent of intravascular inflammatory response. Patients may be quite systemically ill early in the course of severe pneumonia but clinically stabilize later on in empyema development, which should be considered prior to intervention. Patients should always be viewed considering the layers of processes when treating empyema (**Figure 17.1**).


LUNG
Extent of pneumonia
PLEURAL SPACE
Extent of impact on hemithorax
SERUM
Extent of inflammatory response

17.1 Determinants of overall state of illness in patients with empyema.

PREOPERATIVE INVESTIGATIONS AND PREPARATION

The Light criteria for complicated PPE include pH <7.2, lactate dehydrogenase >1000 units, glucose <40 mg/dL or <25% blood glucose, Gram stain or culture positive, and with loculations or septations proven with imaging. A consensus statement from the American College of Chest Physicians in 2000 noted an increase in interventions as the stage of effusion increased. Similarly, a pleural fluid pH less than 7.1 has been found to result in a sixfold increase in the likelihood of surgical intervention based on retrospective data. While these criteria document the physiologic progression of disease, the clinical relevance of the chemical analysis is not critical because the prognostic value is less important in practice. Once pleural space debris causes symptoms and requires removal, drainage is required based on the nature of the debris.

Diagnosis usually follows a progressive clinical picture beginning with pneumonia. Patients with an empyema demonstrate some degree of malaise, persistent fever, respiratory distress, and pleuritic chest pain. Physical exam should exhibit diminished breath sounds with dullness to percussion on the affected side. Ileus, with a lack of appetite, is common.

Initial imaging consists of a chest X-ray (CXR) which shows poor penetration on the affected side. Differentiation of parenchymal consolidation and pleural fluid, however, is difficult using plain radiographs. In a retrospective review of over 300 adult patients, CXR missed all effusions that subsequent computed tomography (CT) scans showed were significant enough to warrant drainage. Decubitus films may be helpful to distinguish between free flowing and loculated effusions.

The next diagnostic step in empyema is an ultrasound or CT scan. Ultrasonography (US) and CT both offer advantages. US is portable, relatively inexpensive, and involves no radiation. It can reliably differentiate between parenchymal and pleural based processes, be used to guide percutaneous drainage and catheter placement, and some authors suggest that US is superior to CT in the identification of pleural debris or loculations. A post hoc review of a prospective trial in children found that, in 31 patients where both CT and US were performed, CT provided no advantage over US in most cases. Two independent series reviewed the implementation of an algorithm using US first in children with complicated pneumonia. Both demonstrated a significant reduction in length of stay and a decrease in the use of CT without an increase in the rate of operative management or pleural drainage.

CT with IV contrast has many of the same advantages as US in that it can differentiate between parenchymal and pleural processes as well as identifying pleural thickening and loculations. However, CT was found to be inferior to US at demonstrating fibrin strands or septations within the pleural fluid. Neither CT nor US is completely reliable in differentiating the specific stages of parapneumonic disease. Consensus statements are also clear in their recommendations of performing CT only when needed, such as for preoperative planning in some cases.

OPERATIONS

Parapneumonic effusion (stages 1–2)

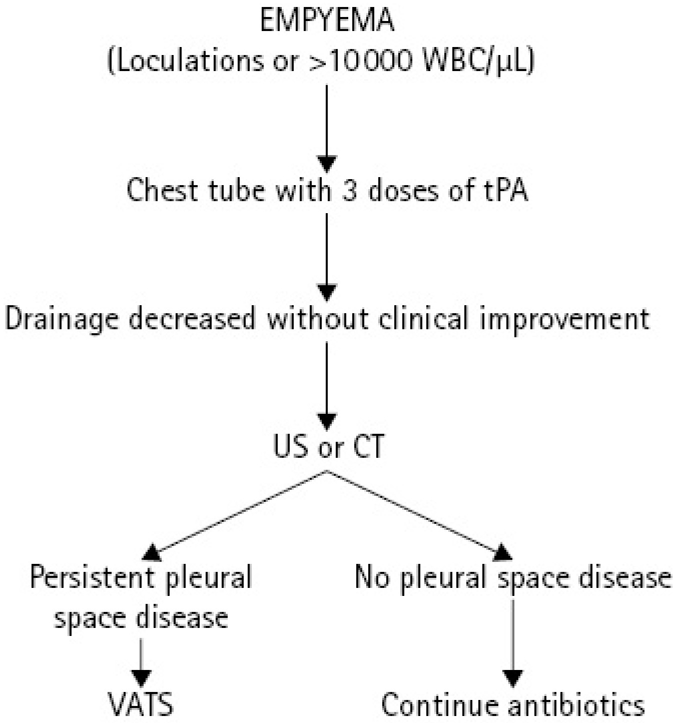
Traditionally, PPEs without solid components are treated by tube thoracostomy and IV antibiotic therapy.

A 12-year retrospective study in children classified small effusions as <1/4 hemithorax opacification, moderate effusions as 1/4–1/2 opacification, and large as >1/2 opacification base on upright films. Most moderate-sized effusions were effectively managed without drainage with no increase in the length of stay or other complications. This suggests that intervention should be based on symptoms, not size alone. Symptoms precipitating intervention are generally poor feeding tolerance, tachypnea, and increasing oxygen requirement.

Upon deciding to drain the fluid, options include single or multiple thoracentesis versus tube thoracostomy. A prospective, non-randomized series compared treating children with repeated US guided needle aspirations versus tube thoracostomy and showed 2.4 drainages/patient with needle aspiration but similar length of stay. While multiple thoracentesis may be reasonable in an older child who could tolerate the procedure with a local anesthetic, it would likely not be appropriate in younger children. The British Thoracic Society guidelines recommend a chest tube for cases in which the first thoracentesis fails to adequately drain the effusion in order to avoid multiple attempts.

On the subject of adequate chest tube size, a series of 405 adult patients confirmed no advantage to tubes larger than 14 Fr. Further, smaller caliber tubes did not hinder the use of fibrinolytics. In a retrospective series of 20 children treated with standard chest tubes compared to 12 treated with pigtail tubes, no differences were found.

In patients with pleural effusions or empyema we use a 12 Fr chest tube (Thal-Quick Chest Tubes, Cook Critical Care, Bloomington, IN) introduced using the Seldinger technique (**Figure 17.2**). We have not felt it necessary to use image guidance in the majority of patients.


EMPYEMA
(Loculations or >10000 WBC/μL)
Chest tube with 3 doses of tPA
Drainage decreased without clinical improvement
US or CT
Persistent pleural space disease
No pleural space disease
VATS
Continue antibiotics

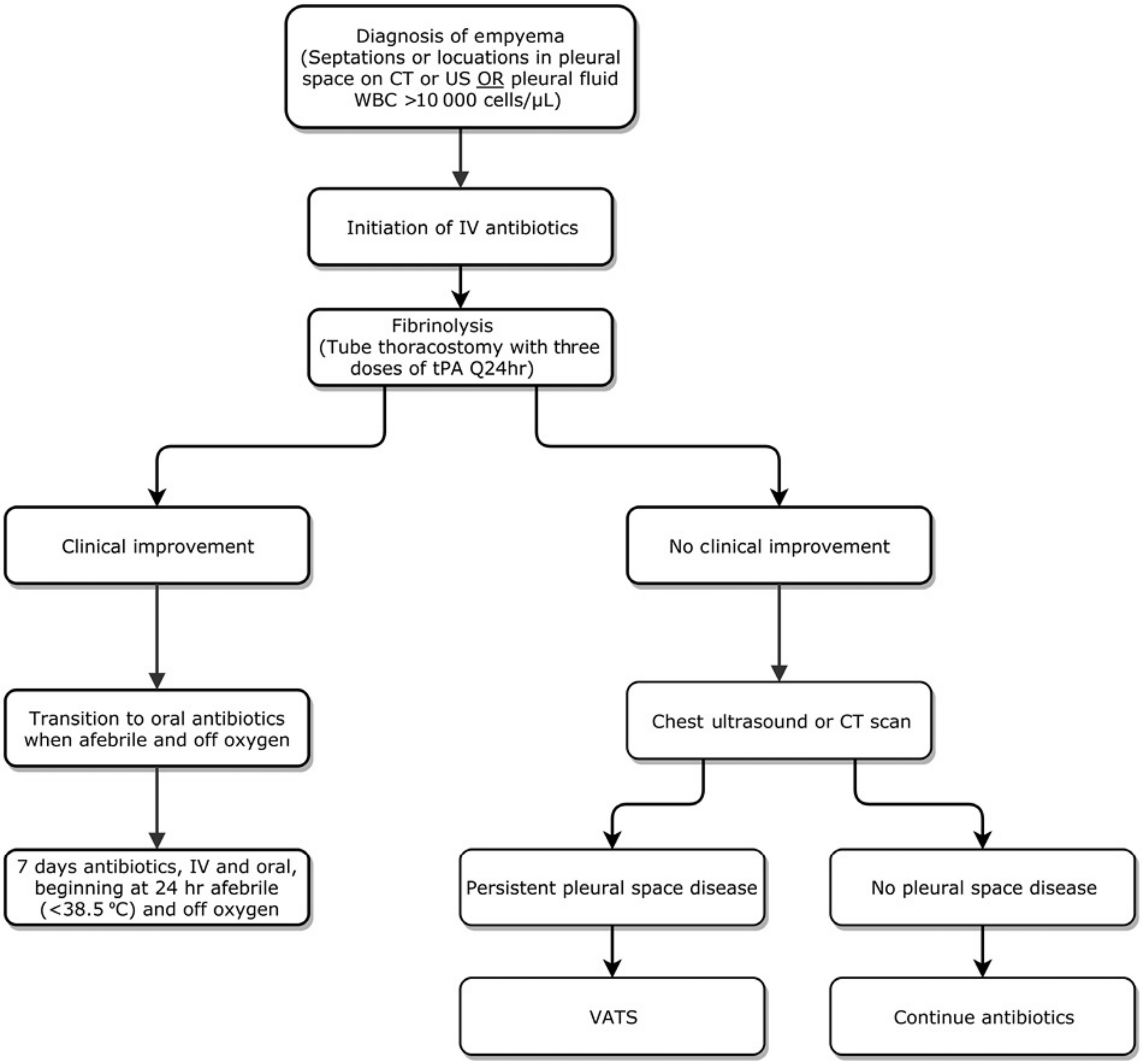
17.2

Empyema (stages 3–4)

The definitive management for empyema has traditionally been surgical debridement, which is currently done via the minimally invasive approach, video-assisted thoracoscopy (VATS). VATS has resulted in earlier and more complete resolution of empyema than chest tube drainage alone in both retrospective and prospective studies, translating to shorter hospitalization with primary VATS. However, primary management of empyema with surgical debridement is changing and fibrinolysis has developed a more prominent role in empyema management.

Three prospective, randomized trials have been conducted independently comparing fibrinolysis to VATS upon diagnosis of empyema in children. The studies were conducted in Spain, the United Kingdom, and the United States in 103, 60, and 36 patients respectively. All three studies compared the installation of fibrinolytic agents for 3 days to VATS as the initial therapy for empyema. The first fibrinolytic dose in both studies was given upon diagnosis and/or chest tube placement. The Spanish study used urokinase every 12 hours for 3 days while the other two studies used 24-hour increments to complete the three-dose course over a 48-hour period. The results were concordant, with all the studies documenting no difference in duration of hospitalization, which was the primary outcome in all the trials. The study conducted in the United States found no difference in days of tube drainage, days of fever, doses of analgesics, or oxygen requirements. The Spanish study also found no difference in time to defervescence. The studies in the United Kingdom and United States documented VATS was more expensive and utilized an intention-to-treat analysis so the length of stay and total charges included the patients who failed fibrinolysis and were subsequently treated with VATS. The failure rate for fibrinolysis was 10% in the Spanish study and 16.6% in the other two studies. These failure rates were similar to previous studies investigating the utility of fibrinolysis. In addition to the pediatric trials, a meta-analysis of 10 trials, including adult studies with over 900 patients, documented similar conclusions as the pediatric trials, that fibrinolysis is an effective first-line therapy.

An algorithmic approach to patients with empyema is employed and summarized in **Figure 17.3**. The critical consideration prior to embarking on thoracoscopic or open debridement for empyema is the status of the underlying parenchyma. Patients with persistent illness may have pulmonary necrosis as their source instead of pleural space disease. In this circumstance, an operation should be avoided in favor of patience.


Diagnosis of empyema (Septations or locuations in pleural space on CT or US OR pleural fluid WBC >10 000 cells/µL)
Initiation of IV antibiotics
Fibrinolysis (Tube thoracostomy with three doses of tPA Q24hr)
Clinical improvement
No clinical improvement
Transition to oral antibiotics when afebrile and off oxygen
7 days antibiotics, IV and oral, beginning at 24 hr afebrile (<38.5 °C) and off oxygen
Chest ultrasound or CT scan
Persistent pleural space disease
No pleural space disease
VATS
Continue antibiotics

17.3 An algorithmic approach to patients with empyema. Either a US or CT scan is performed to evaluate the extent of disease. Tube thoracostomy is employed with three subsequent daily doses of tPA given. If, after three doses, the patient has not clinically improved, another US or CT scan is performed to evaluate for intrapleural disease. Minimally invasive pleural debridement (i.e. VATS) is considered eventually if the tPA has failed to eradicate the pleural space of fibrinopurulent material.

Tube thoracostomy and pleural fibrinolysis

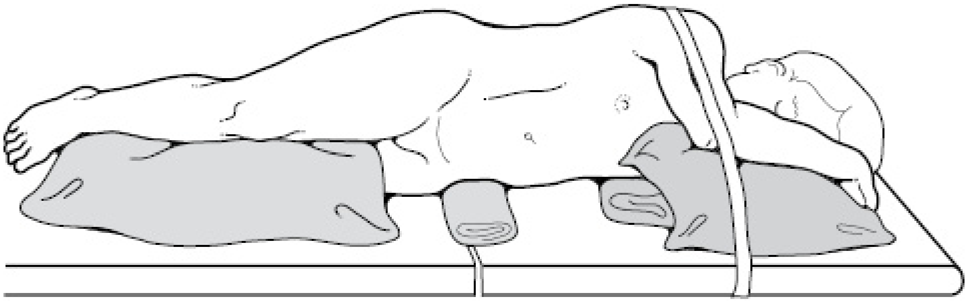
Patients are sedated according to local standards. The skin and soft tissues about the fifth intercostal space in the mid-axillary are infiltrated with either 1/4% marcaine or 1% lidocaine (1 mL/kg maximum). For instillation of fibrinolytics, we prefer to use a 12 Fr chest tube (Thal-Quick Chest Tubes, Cook Critical Care, Bloomington, IN) introduced using the Seldinger technique. We have not felt it necessary to use image guidance in the majority of patients. Once the tube is inserted, the effluent is sent for culture, and the tube is secured to a −20 cm $H_2 0$ suction collection device.

Fibrinolysis is performed by mixing 4 mg of tissue plasminogen activator (tPA) in 40 mL of normal saline. The prepared tPA solution is injected directly into the tube, which is subsequently clamped for 1 hour. After the dwell time, the tube is returned to continuous suction. Two additional doses are subsequently given at 24-hour intervals utilizing the same technique. The tube is removed once there is no evidence of an air leak and the output is less than 1 mL/kg/day.

We no longer place traditional chest tubes for the treatment of empyema. Alternatives include the use of a 10 mm expandable sleeve and cannula (Step□, Covidien Surgical, Mansfield, MA, USA) as an avenue for insertion of a soft-suction drain (19 Fr round BLAKE□ drain), or a 5 mm port for a smaller drain.

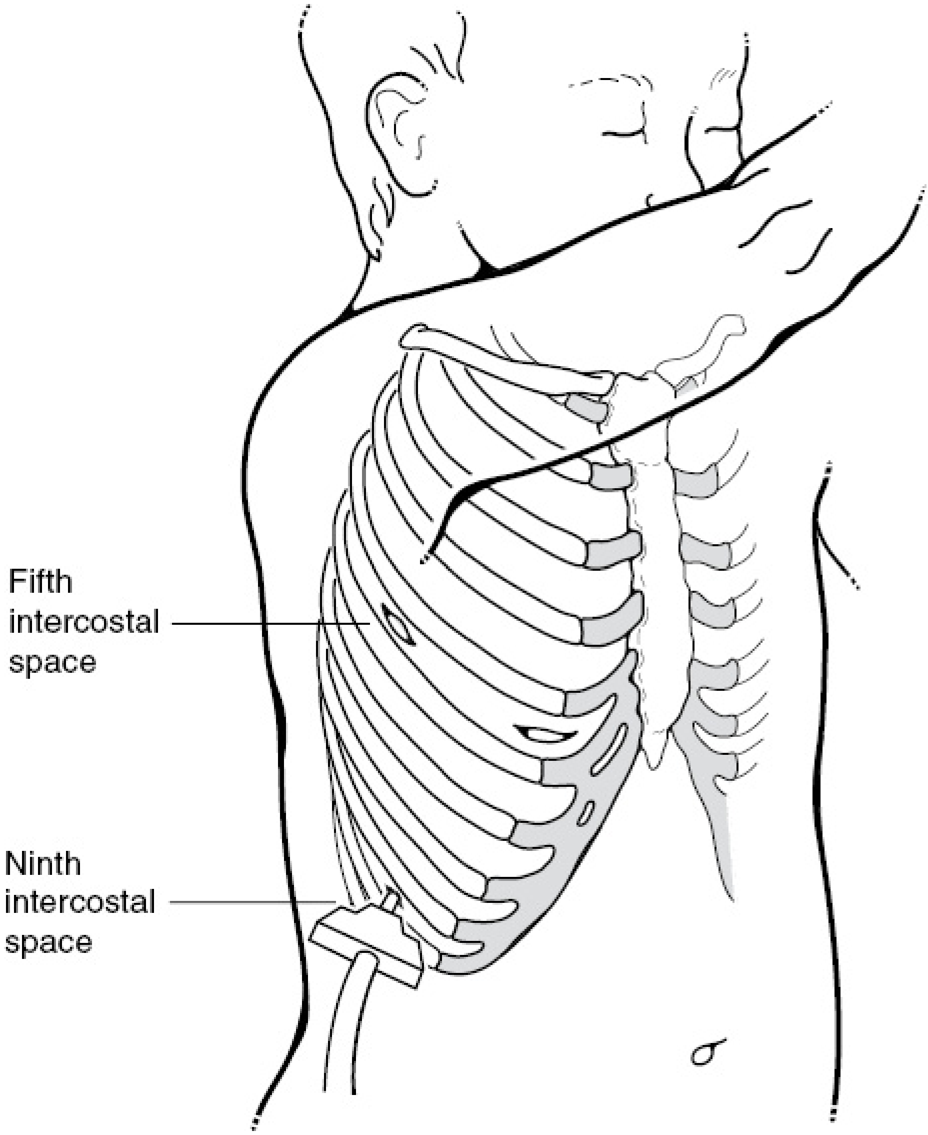
Video-assisted thoracoscopic debridement

For each of the following operative techniques, the procedure is performed under general anesthesia with tracheal intubation and is facilitated by selective, single-lung ventilation. The patient is positioned in the lateral decubitus position with the affected side up, and an axillary roll is placed (**Figure 17.4**). One should aim to position the patient with the level of the iliac crest resting at the break in the table to allow for increased opening of the intercostal spaces. Raising the kidney rest, if available, can also assist in this goal.



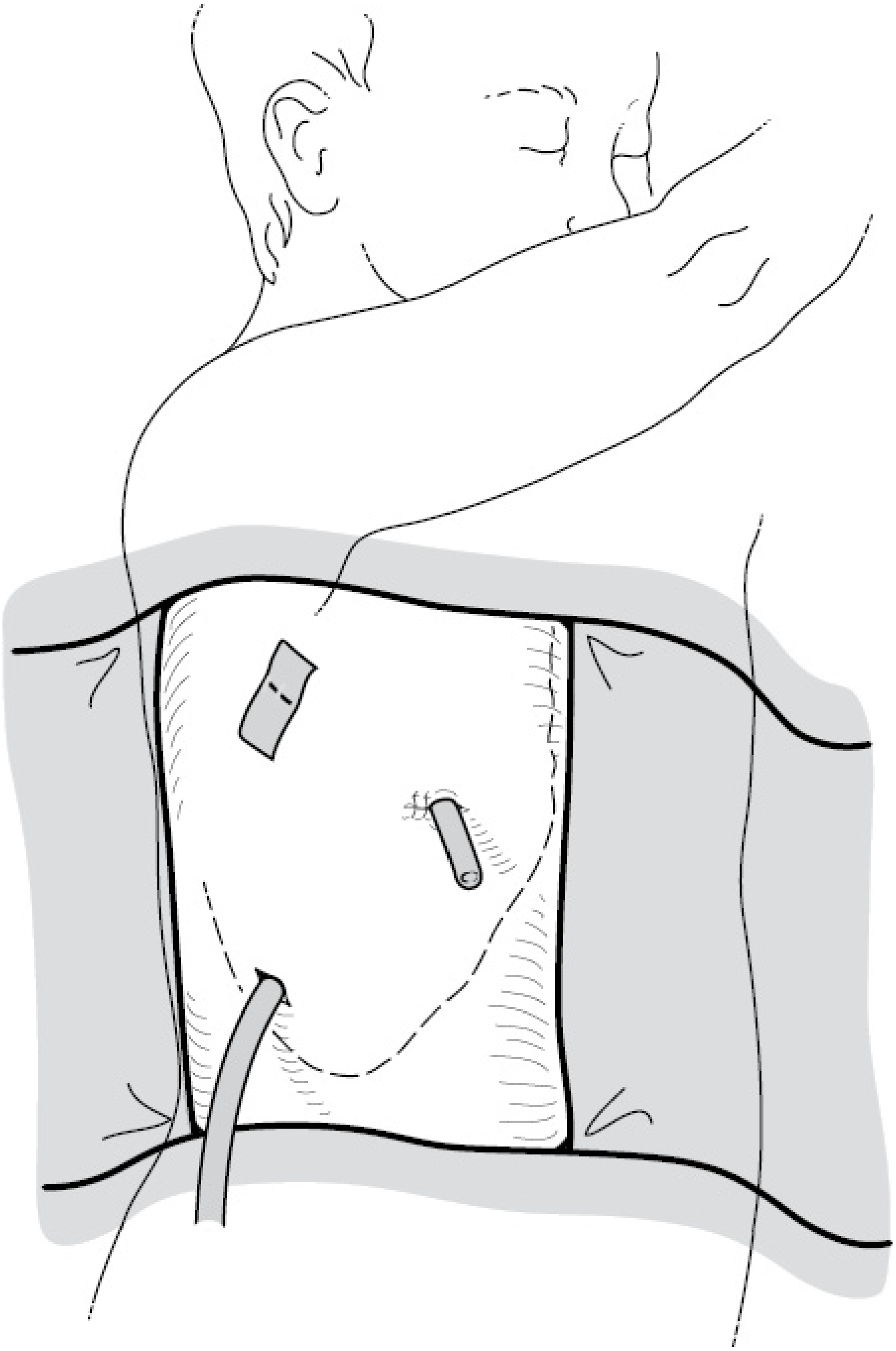
17.4

The initial incision and insertion of a 10 mm port is positioned in the area deemed advantageous for accessing the majority of disease after review of the US or CT scan. Typically, this initial 10 mm port is best placed directly overlying the empyema. Placement of the port in the fifth intercostal space at the mid-axillary line is a good starting point (**Figure 17.5**). A low pressure (4 mmHg), low flow (1 L/min) CO_2 pneumothorax facilitates lung collapse. A 10 mm angled telescope is used to create a working space by sweeping the adhesions and lung away from the chest wall with the end of the telescope. Once the underlying lung is freed adequately, a second incision is placed in the location that most easily allows for the majority of pleural disease to be removed. When two incisions are not sufficient, a small third incision for a 5 mm port can be placed to provide a second working port. Pleural debridement is then undertaken using a ring forceps or Yankauer suction to remove the pleural peel and debris. Alternatively, an atraumatic grasping forceps can also be used. Samples for Gram stain and culture are routinely sent by attaching a Lukens trap to the suction device. Once all pockets of fluid have been drained and the majority of the pleural debris is removed, a single chest drain in the form of a soft, flexible drain (preferably a 19 Fr round Blake drain, if available) is inserted into the pleural space under direct vision.


Fifth intercostal space
Ninth intercostal space

17.5

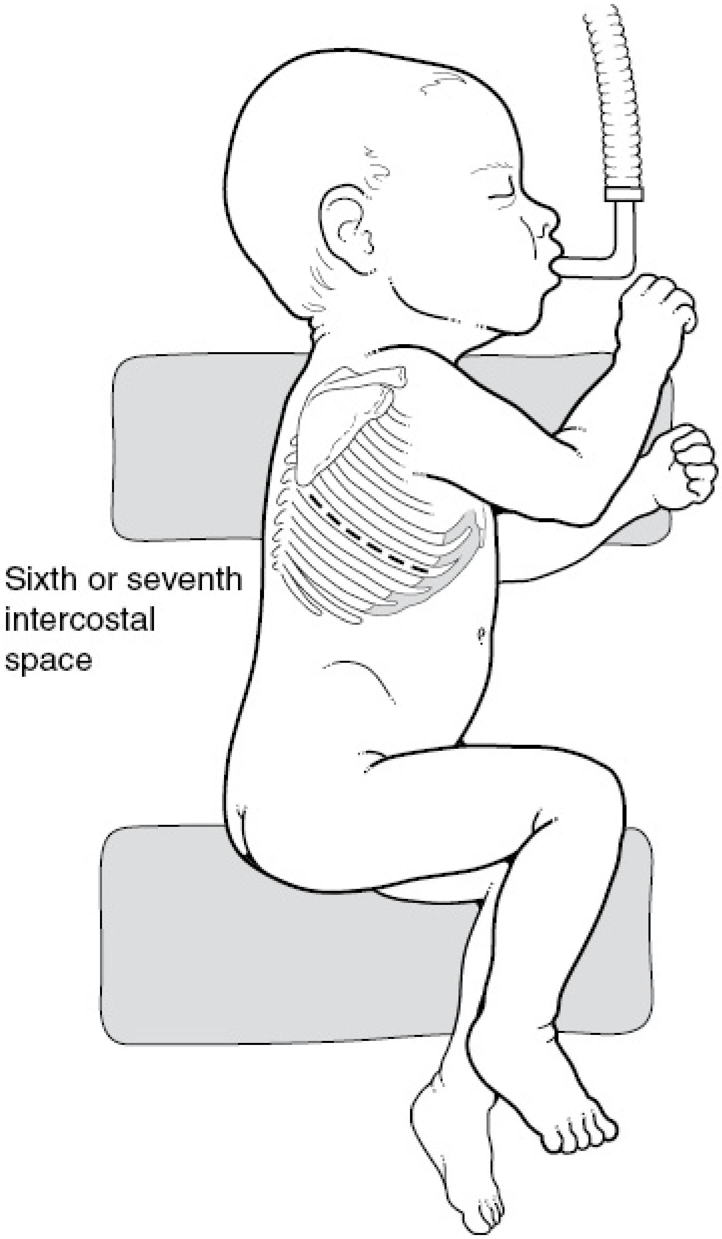
To lessen the possibility for pneumothorax when the tube is removed, the drain is tunneled over the rib space cephalad to the port site through which it is going to exit. Incisions are closed with absorbable suture and sterile dressings are applied (**Figure 17.6**).



17.6

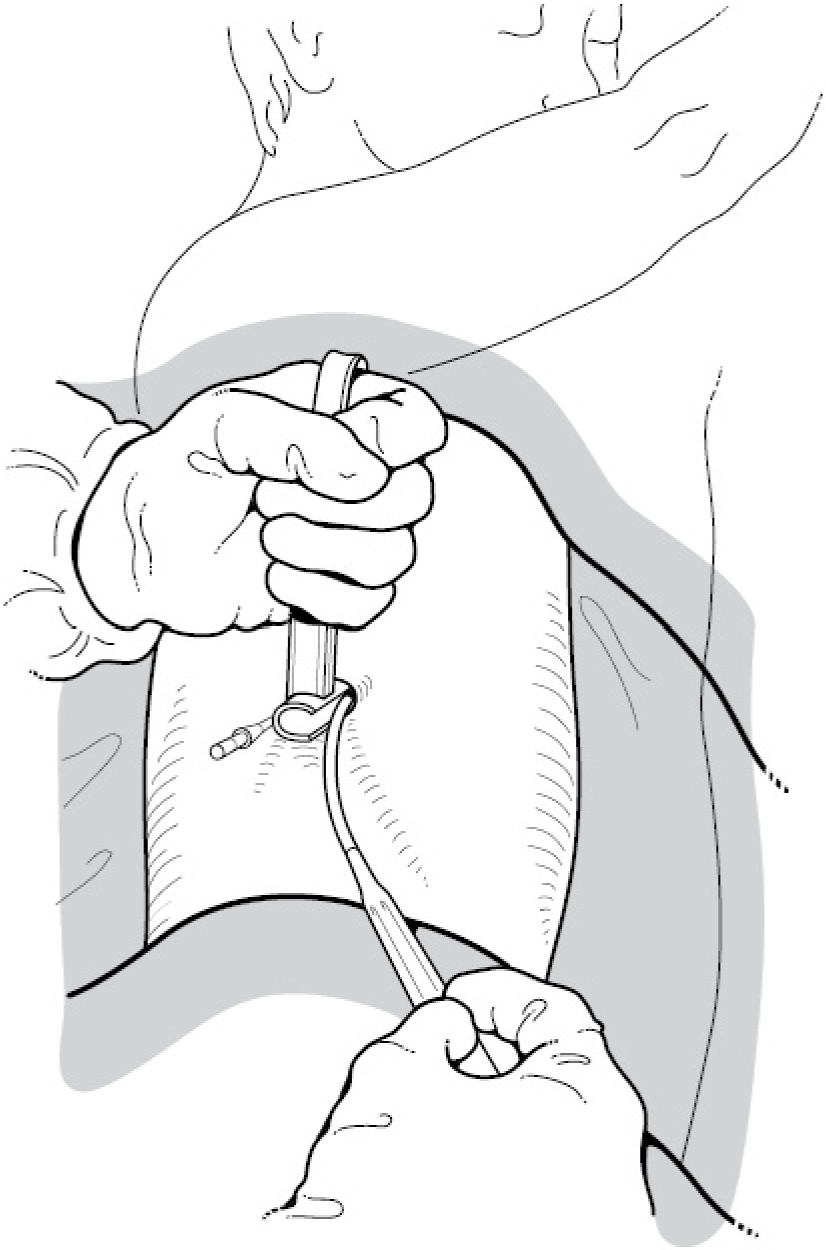
Mini thoracotomy pleural debridement

When videoscopic equipment is not available, pleural debridement through a small thoracotomy incision can be easily performed. In addition, thoracoscopic debridement in small children can be accomplished via mini-thoracotomy with equivalent results. A small (3 cm) incision is situated in the mid-axillary line at the level of the fifth intercostal space (**Figure 17.7**).



17.7

Use of an appropriately sized mediastinoscope with a good light source facilitates visualization of the pleural space (**Figure 17.8**). Debridement ensues under direct vision. A suction device can be inserted through the mini-thoracotomy whereby loculations and debris are bluntly removed and lysed. An appropriate-sized chest drain is tunneled over the ribs caudal to the incision. The intercostal muscles are reapproximated with polygalactin suture and the chest drain is secured to the skin with non-absorbable suture.



17.8

Thoracotomy

Pleural debridement via a posterolateral thoracotomy is reserved for patients in whom thoracoscopy or mini-thoracotomy is unsafe or has proven ineffective. However, this is almost never required in children.

POSTOPERATIVE CARE

The chest drain is initially placed to a pleurovac with 20 cm of H_2O suction. If there is no air leak or the output is not severe, this can be transitioned to a bulb suction. A postoperative chest radiograph in the recovery room is not necessary. The chest drain is maintained on suction until drainage is less than 1 mL/kg/day in a 24-hour period. The drain can be pulled at that point without the need for post-pull X-ray unless otherwise indicated.

The current consensus guidelines on empyema recommend antibiotics 10 days after resolution of fever. This is expert opinion only and emerging data suggest 7 days may be adequate. Patients are discharged home when the chest drain has been removed, pain is controlled, and nutritional issues are optimized. A follow-up visit and chest radiograph are recommended 3–4 weeks following discharge.

OUTCOME

The results from the prospective randomized controlled trials showed no significant difference in several important outcome variables including length of stay, cost, and resolution of fever between initial thoracoscopic debridement and decortication (TDD) and intrapleural fibrinolytic therapy. Although we advocate an initial attempt at fibrinolytic therapy in patients with empyema, we recognize that primary TDD is likely equally as effective given the circumstances of one's practice. Despite the availability of Level I evidence to guide therapy, these decisions will continue to be governed by historical and practice-specific algorithms.

FURTHER READING

Bradley JS, Byington CL, Shah SS, et al. Executive summary: The management of community acquired pneumonia in infants and children older than 3 months of age: Clinical practice guidelines by the Pediatric Infectious Disease Society and the Infectious Disease Society of America. *Clin Infect Dis* 2011; 53: 617–30.

Islam S, Calkins CM, Goldin AB, et al. The diagnosis and management of empyema in children: A comprehensive review from the APSA Outcomes and Clinical Trials Committee. *J Pediatr Surg* 2012; 47(11): 2101–10.

Kurt BA, Winterhalter KM, Connors RH, et al. Therapy of parapneumonic effusions in children: video-assisted thoracoscopic surgery versus conventional thoracostomy drainage. *Pediatrics* 2006; 118(3): e547–53.

Marhuenda C, Barceló C, Fuentes I, et al. Urokinase versus VATS for treatment of empyema: A randomized multicenter clinical trial. *Pediatrics* 2014; 134(5): e1301–7.

Singh M, Mathew JL, Chandra S, et al. Randomized controlled trial of intrapleural streptokinase in empyema thoracis in children. *Acta Paediatr* 2004; 93(11): 1443–5.

St Peter SD, Tsao K, Spilde TL, et al. Thoracoscopic decortication vs tube thoracostomy with fibrinolysis for empyema in children: A prospective, randomized trial. *J Pediatr Surg* 2009; 44(1): 106–11.

Sonnappa S, Cohen G, Owens CM, et al. Comparison of urokinase and video-assisted thoracoscopic surgery for treatment of childhood empyema. *Am J Respir Crit Care Med* 2006; 174(2): 221–7.

Valusek PA, Tsao K, St. Peter SD, et al. A comparison of chest tubes versus bulb-suction drains in pediatric thoracic surgery. *J Pediatr Surg* 2007; 42(5): 812–14.

Velaiutham S, Pathmanathan S, Whitehead B, Kumar R. Video-assisted thoracoscopic surgery of childhood empyema: Early referral improves outcome. *Pediatr Surg Int* 2010; 26(10): 1031–5.

18

Chylothorax

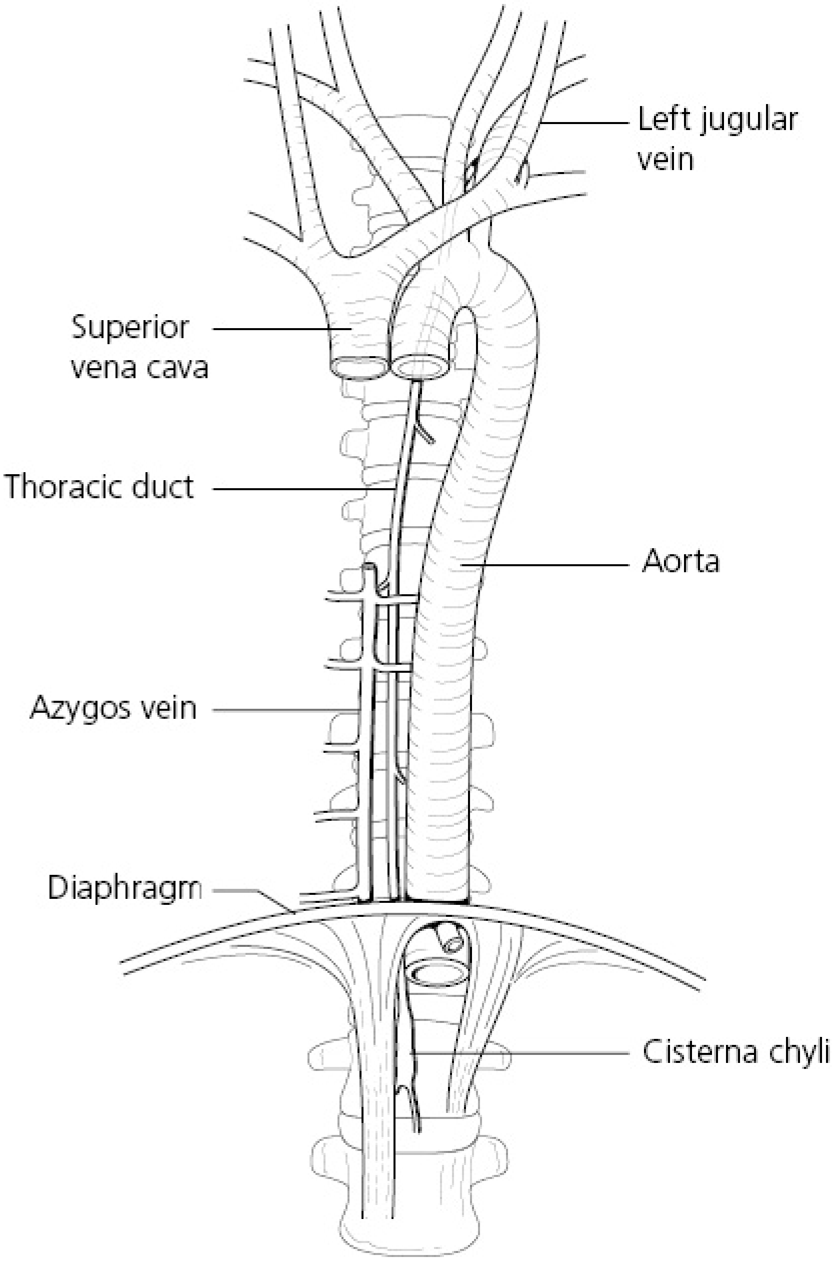
SHAUN M. KUNISAKI

INTRODUCTION

A chylothorax is a pleural effusion composed of lymphatic fluid. In children, the most common etiology is iatrogenic following an operation within the posterior mediastinum. Procedures on the esophagus and thoracic aorta, such as ligation of a patent ductus arteriosus, coarctation repair, and Fontan procedure, are at highest risk for this complication. Ten per cent of chylothoraces in the pediatric population are congenital. Other important causes of chylothoraces in children include superior vena cava obstruction, blunt and penetrating trauma, lymphatic malformations, malignancy, and following diaphragmatic hernia repair, particularly in the setting of right heart failure.

PRINCIPLES AND JUSTIFICATION

The thoracic duct, which carries 70–90% of all fats absorbed by the gut into the venous circulation, originates within the abdomen at the cisterna chyli located over the second lumbar vertebral body (**Figure 18.1**). The duct extends into the thorax through the aortic hiatus and then passes upward into the posterior mediastinum on the right before shifting toward the left at the level of the fourth or fifth thoracic vertebra. It then ascends behind the aortic arch and into the posterior neck to the junction of the left subclavian and internal jugular veins. Therefore, disruption of the thoracic duct below the fifth thoracic vertebra produces a right chylothorax. Disruption of the thoracic duct above the fifth thoracic vertebra produces a left chylothorax.


Left jugular vein
Superior vena cava
Thoracic duct
Aorta
Azygos vein
Diaphragm
Cisterna chyli

18.1

Any child with the suspicion of having a chylothorax mandates an analysis of the pleural fluid obtained by thoracentesis or tube thoracostomy. Chylous fluid appears straw colored in chronically fasted patients but turns milky with enteral feeding. A pleural fluid cell count greater than 1000 cells per microliter with more than 70% lymphocytes is diagnostic for a chylothorax. In older non-fasted children, a triglyceride level of more than 110 mg per deciliter supports the diagnosis.

A trial of medical management is always indicated before operative management for a chylothorax is attempted. All patients require an indwelling transthoracic chest tube (12 Fr) to drain the fluid and to promote pulmonary re-expansion. Because of the small caliber of these tubes, they may require stripping several times daily to ensure patency. Aside from the obvious respiratory embarrassment from the accumulation of chyle in the pleural space, a chylothorax, even in small infants, can produce several hundred milliliters of fluid per day, resulting in hyponatremia, hypoalbuminemia, and immunologic derangements secondary to the loss of lymphocytes. The latter can lead to an overwhelming bacterial and/or fungal sepsis and death. Serum electrolytes, complete blood counts including absolute lymphocytes, coagulation, albumin, and total IgG levels should be monitored at least once weekly. Electrolyte derangements and intravascular volume losses should be corrected with crystalloid or 5% albumin. The use of prophylactic antibiotics has not been shown to be effective.

A major cornerstone in the initial treatment of any chylothorax is complete restriction of enteral intake with the administration of IV alimentation for a minimum of 7 days. In the setting of continued high output (>20 mL/kg/day) drainage, continuous IV somatostatin therapy (initial dose, 3 mcg/kg/hour) has been used as an adjunct to facilitate closure of the leak, but its efficacy is questionable. Somatostatin is thought to act on gastric, pancreatic, and biliary secretions to decrease the overall volume and protein content of fluid going to the thoracic duct. Serum glucose and thyroid hormone levels need to be monitored during somatostatin therapy. Finally, high clinical suspicion for central venous obstruction as an underlying cause for the chylothorax should be evaluated by magnetic resonance venography since such cases may be amenable to IV thrombolysis and/or intravascular balloon dilation. If the chest tube output improves and there is a minimal effusion by chest ultrasound, the child can be advanced to a high medium-chain triglycerides enteral diet for 4–6 weeks before transitioning to a regular diet.

If these initial management strategies fail after 1–4 weeks, operative management is indicated. The exact timing of surgical intervention depends on the etiology, volume of output, severity of nutritional/immunologic depletion, and clinical stability of the patient. Preoperative lymphoangiograms or nuclear scintigraphy can be performed, but these studies are technically difficult to obtain and are seldom helpful in operative planning.

ANESTHESIA

General anesthesia is usually most appropriate although pleuroperitoneal shunts can be placed under local anesthesia, if desired. Lung isolation is ideal in thoracoscopy and thoracotomy because of the need to clearly visualize the medial aspect of the posterior mediastinum. This can usually be accomplished by selective intubation of the contralateral mainstem bronchus. In toddlers and school-age children, a bronchial blocker is another good option. Adolescents should receive a double-lumen endotracheal tube.

The management of postoperative pain should be discussed with the anesthesiologist prior to the operation. Options include patient- or nurse-controlled anesthesia, thoracic epidural catheters, and intermittent IV dosing, depending on whether a thoracotomy is planned and the age of the patient.

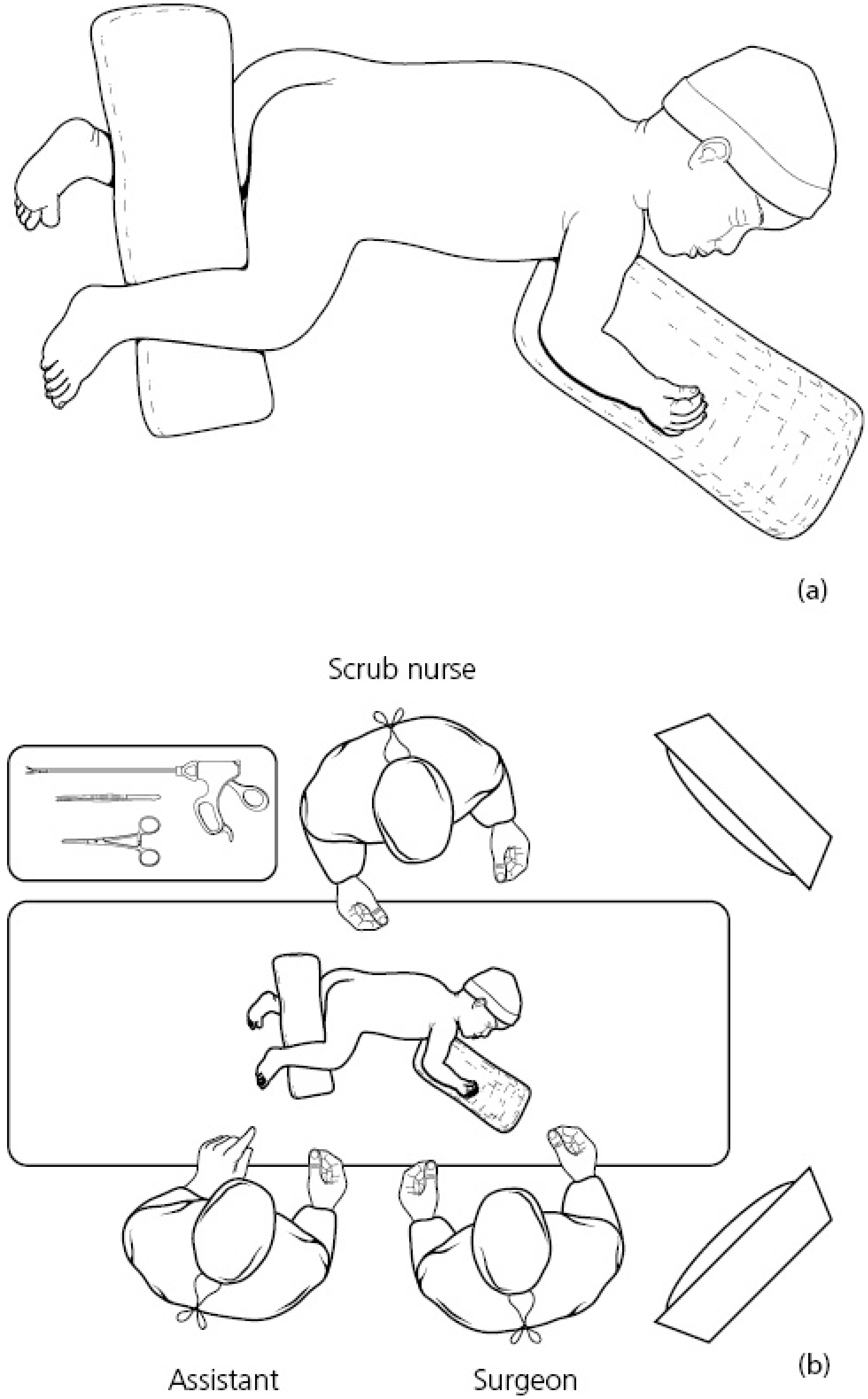
OPERATIONS

The surgical armamentarium for the management of refractory chylothoraces includes thoracic duct ligation, pleurodesis, pleurectomy, and pleuroperitoneal shunts. Each of these procedures can be performed as a stand-alone procedure or in combination, depending on surgeon preference and the clinical scenario. Percutaneous thoracic embolization, performed by a skilled interventional radiologist, is an emerging alternative to operative to surgical thoracic duct ligation that warrants further study.

Thoracic duct ligation

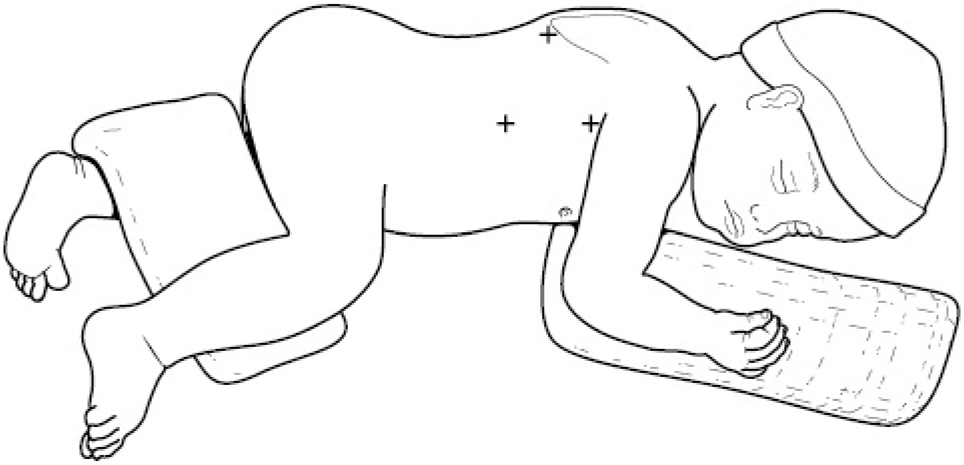
Thoracic duct ligation is the preferred operation in suspected cases of localized, iatrogenic injury. The thoracic duct should be approached from the ipsilateral side, whereas in cases of bilateral effusion, the right chest should be chosen initially. Thoracoscopic duct ligation has become the current standard given its superior view of the posterior mediastinum at the level of the diaphragm, even in neonates and small infants. However, some children, such as those with significant pulmonary adhesions or severe congenital diaphragmatic hernia, should be approached via a posterolateral thoracotomy, typically through the sixth or seventh interspace.

One hour prior to operative incision, approximately 60 mL of heavy cream should be administered through a post-pyloric feeding tube. This maneuver can occasionally be helpful in identifying the chyle leak intraoperatively. Because of the post-pyloric position, infusion may continue during the procedure, itself; and this may increase the likelihood of detection of a thoracic duct leak. The child is positioned in a modified prone position with 45 degrees of elevation of the ipsilateral thorax with padded rolls or a beanbag (**Figure 18.2a and b**). An axillary roll is placed to minimize the risk of brachial plexus injury. The ipsilateral arm is elevated anteriorly out of the way or alternatively prepped in its entirety into the sterile field.


(a)
Scrub nurse
Assistant
Surgeon
(b)

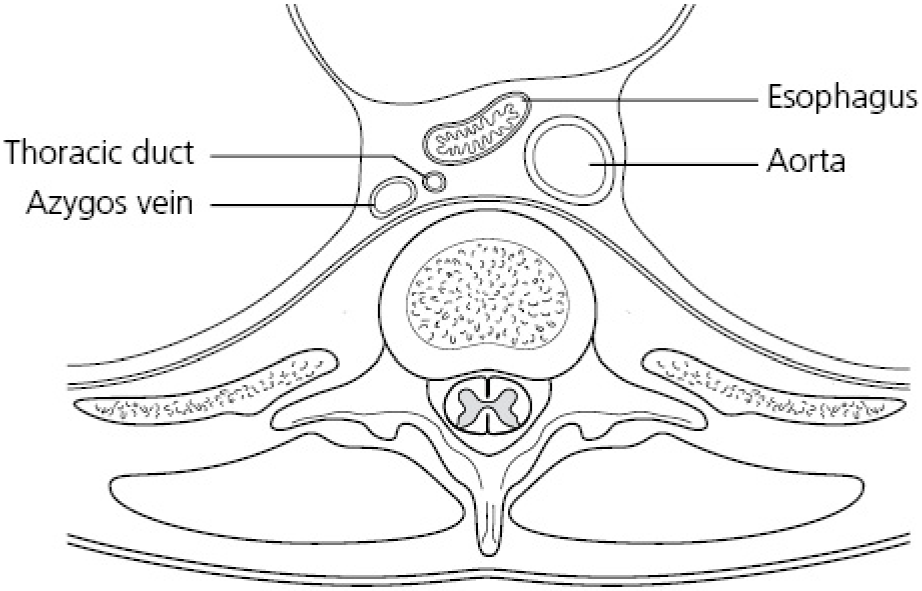
18.2a,b

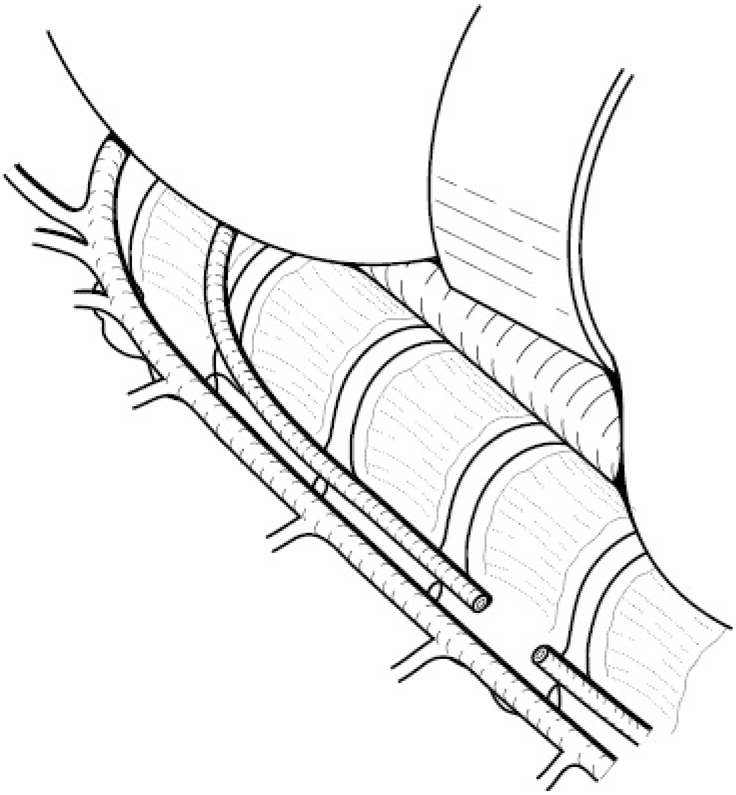
A 5 mm incision is made in the sixth intercostal space midway between the nipple and scapula tip along the mid-axillary line. The pleural space is entered with a Veress needle or artery forceps. A 5 mm port is placed, and the hemithorax is insufflated with carbon dioxide to 4 mmHg. The posterior mediastinum is visualized using a 5 mm 30-degree thoracoscope. Two additional ports are placed in a triangular configuration along the mid-axillary and posterior axillary lines (**Figure 18.3**). This orientation of the ports allows for optimal visualization of the posterior mediastinum at the level of the diaphragm.



18.3

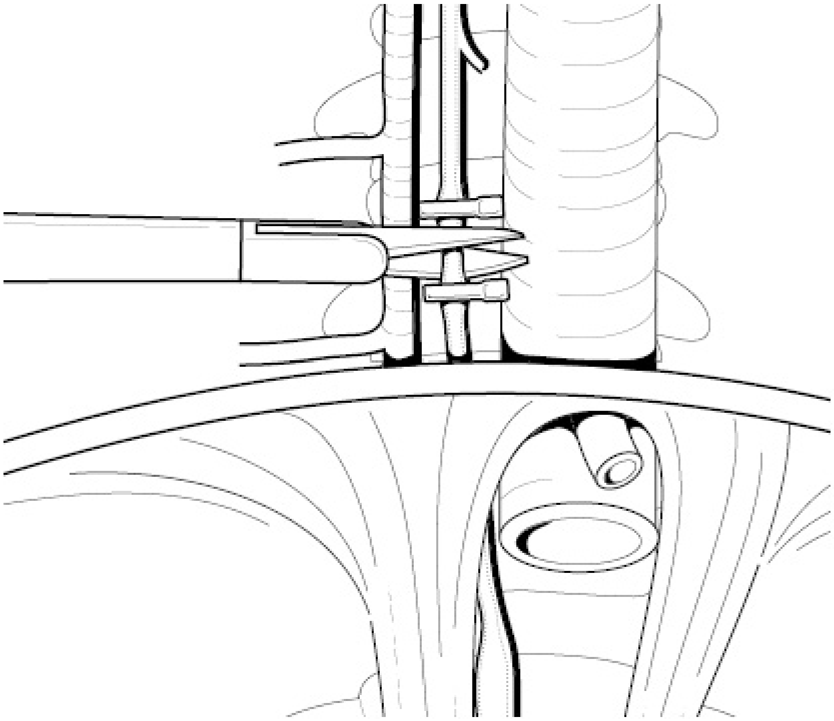
Depending on the size of the patient, either 3 mm or 5 mm instruments are used to mobilize the lung and take down any adhesions. The inferior pulmonary ligament can be divided with diathermy, if necessary, to optimize exposure. The esophagus is visualized with the assistance of an orogastric tube placed by the anesthesiologist at induction. On the right side, the thoracic duct is located between the azygous vein and the aorta (**Figure 18.4**).


Esophagus
Thoracic duct
Aorta
Azygos vein



18.4

Once the duct is identified, it should be carefully ligated as it enters the chest through the aortic hiatus with small metallic endoscopic clips (**Figure 18.5**). This should be performed regardless of whether a specific area of leakage is identified. The duct can similarly be ligated cephalad to the area of leak site if one is identified. The duct can also be ligated using vessel sealing technology. This is especially true in smaller patients who may not accommodate a 5 mm clip well. Another technique, especially if a definitive duct is not visualized, is a figure-of-eight ligature of the tissue between the azygos vein and esophagus using fine absorbable sutures. If the duct is ligated properly, the surgeon will see an immediate cessation of pleural fluid accumulation. After ligation, it is often advantageous to cover the area of the thoracic duct with fibrin glue or a pleural flap. At the conclusion of the procedure, a chest tube is placed through the inferior port site.



18.5

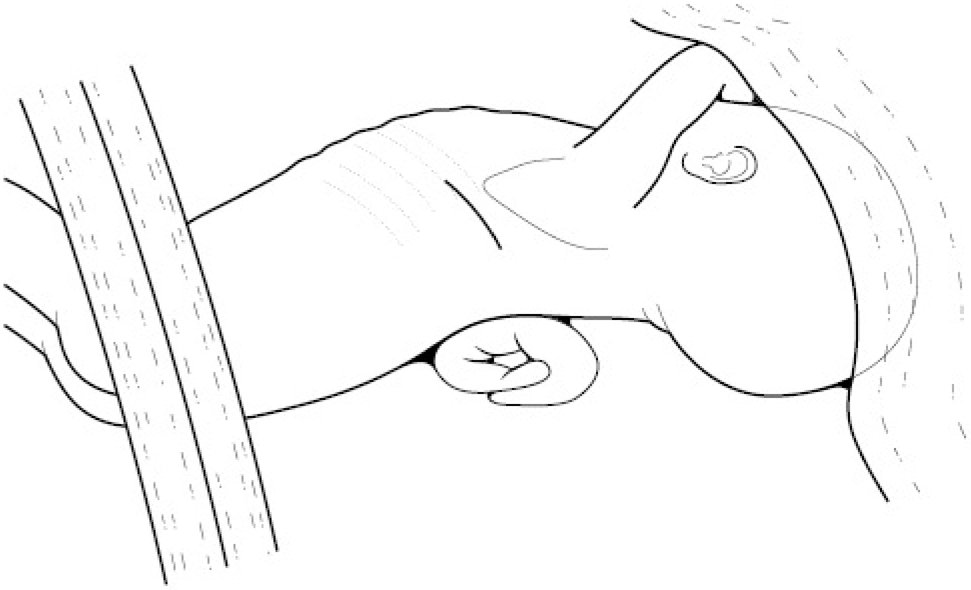
Pleurodesis

Although pleurodesis can be performed as a stand-alone operation to treat a refractory chylothorax, the procedure is more commonly employed in combination with thoracic duct ligation, particularly if no focal leak is identified. Once access to the parietal pleura is obtained either by thoracotomy or thoracoscopy, the entire parietal pleura of the lower thorax adjacent to the thoracic duct is mechanically abraded with a surgical sponge, peanut, or cautery scratch pad. An alternative approach to pleurodesis is to instill a chemical agent such as bleomycin, doxycycline, or talc, all of which are known to facilitate significant pleural inflammation. Chemical pleurodesis is generally reserved for treatment in older children and in patients with congenital lymphangiomatosis. A chest tube should be left in place after pleurodesis.

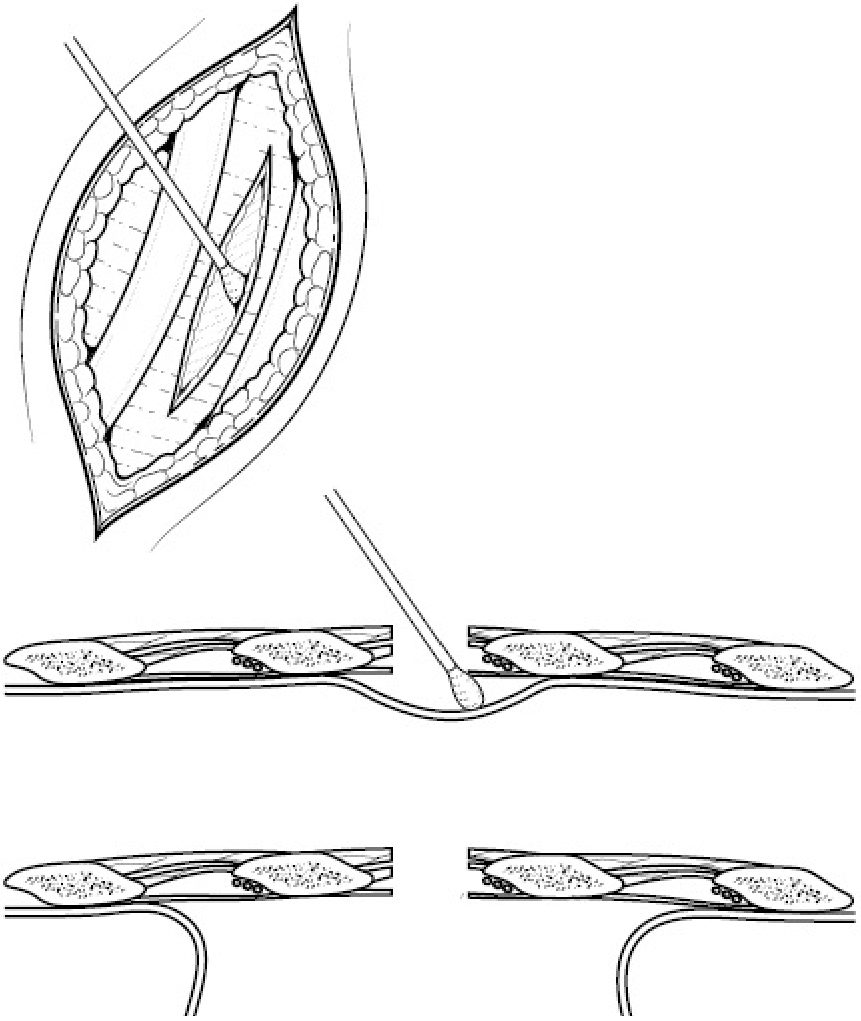
Parietal pleurectomy

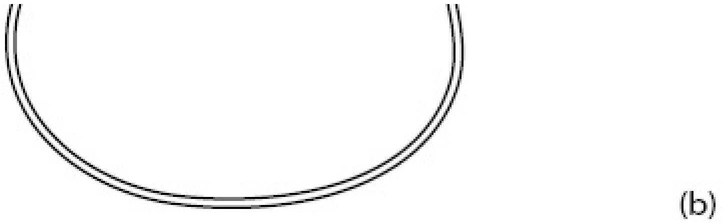
Pleurectomy, which involves manual stripping of full-thickness parietal pleural, is a preferred technique at many institutions in the setting of chylothorax secondary to central venous obstruction or diffuse lymphatic leak.

Pleurectomy is routinely performed through a fifth or sixth interspace posterolateral thoracotomy (**Figure 18.6a**). The posterior mediastinum is accessed using an extrapleural dissection with the aid of cotton-tipped applicators or Freer elevator (**Figure 18.6b**). Once the thoracic duct is exposed, the entire parietal pleura adjacent to the diaphragm and lung hilum is mobilized and excised from the chest wall with diathermy. Special attention is paid toward avoiding injury to the phrenic nerve medially and the recurrent laryngeal nerve superiorly. Although the procedure is rarely associated with significant blood loss, diffuse oozing from the chest wall surfaces may occur in cases where a previous thoracotomy has been performed. A relatively large bore chest tube should therefore be left *in situ*. Pleurectomy may be particularly useful in cases where the leak is diffuse, or is poorly identified (e.g. superior vena caval occlusion).



(a)



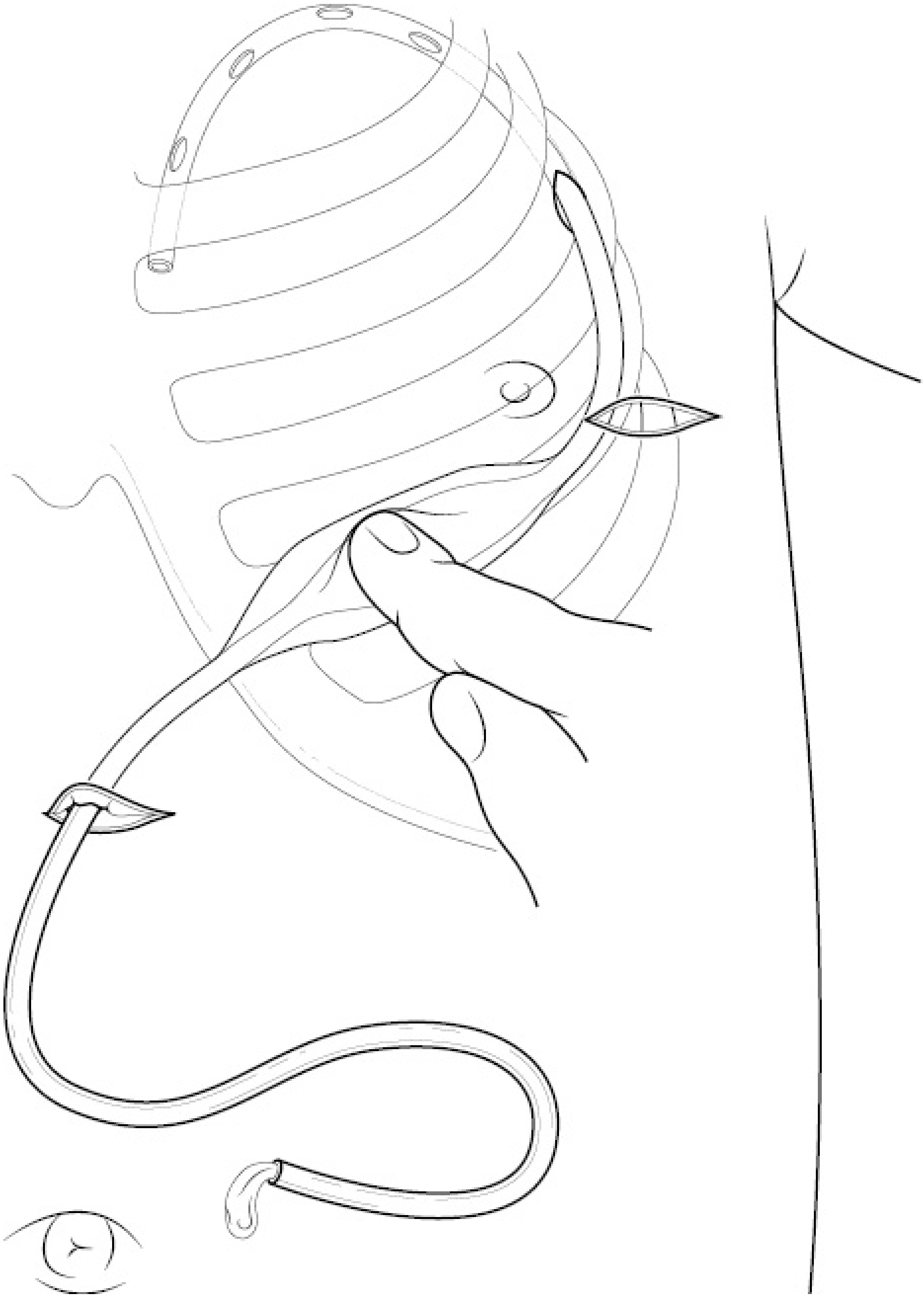


18.6a,b

Pleuroperitoneal shunts

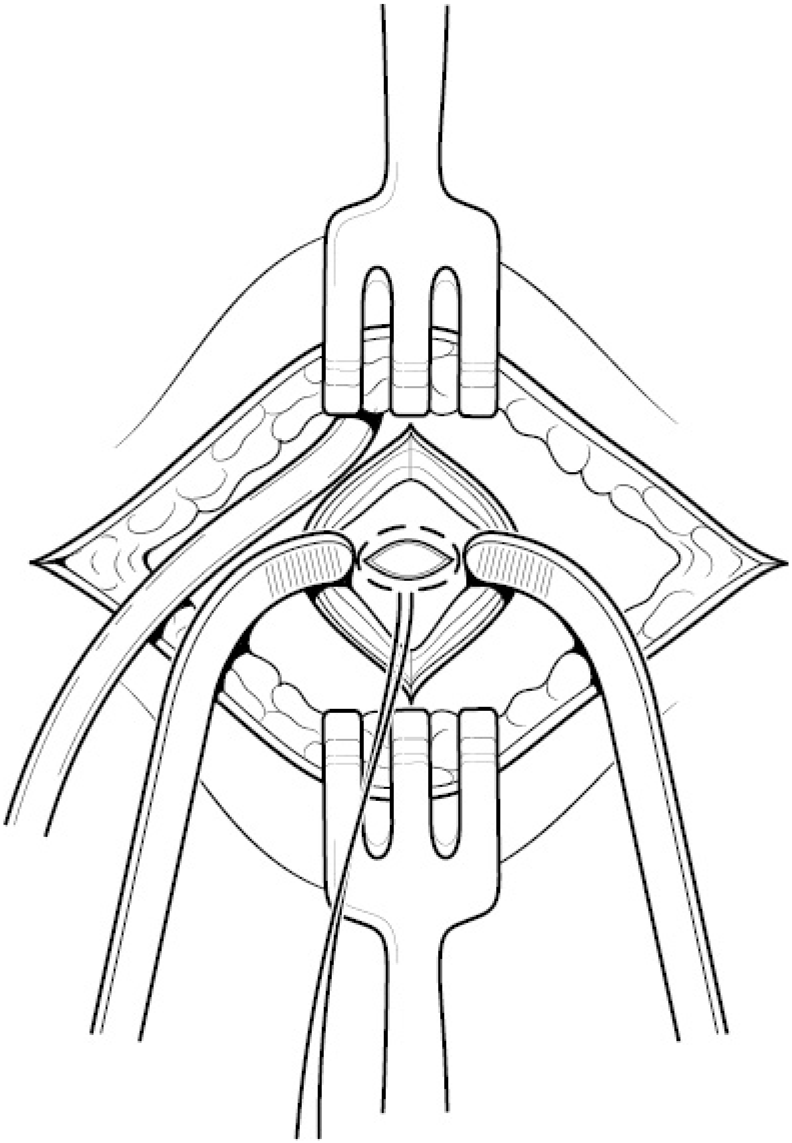
The placement of a shunt is another potential option in the treatment of chylothorax. These shunts are less invasive, making them ideal for high operative risk patients. However, shunts are associated with relatively high failure rates, particularly in infants, and are therefore reserved as a last resort in those who have failed other operative interventions. The one-way pumping chamber can be positioned either externally or internally in the subcutaneous tissues depending on the size of the patient. Although peritoneovenous shunts can also be used for the management of chylothoraces, pleuroperitoneal shunts are more popular and are well described in the literature.

A small transverse skin incision is made along the mid-axillary line caudal to the proposed intercostal space for pleural catheter insertion. A second small skin incision is made over the anterior rectus sheath midway between the umbilicus and xiphoid (**Figure 18.7**). The fibers of the rectus sheath are separated longitudinally. Two concentric purse-string absorbable sutures are placed through the posterior rectus sheath and peritoneum. A subcutaneous tunnel between the two incisions is created.



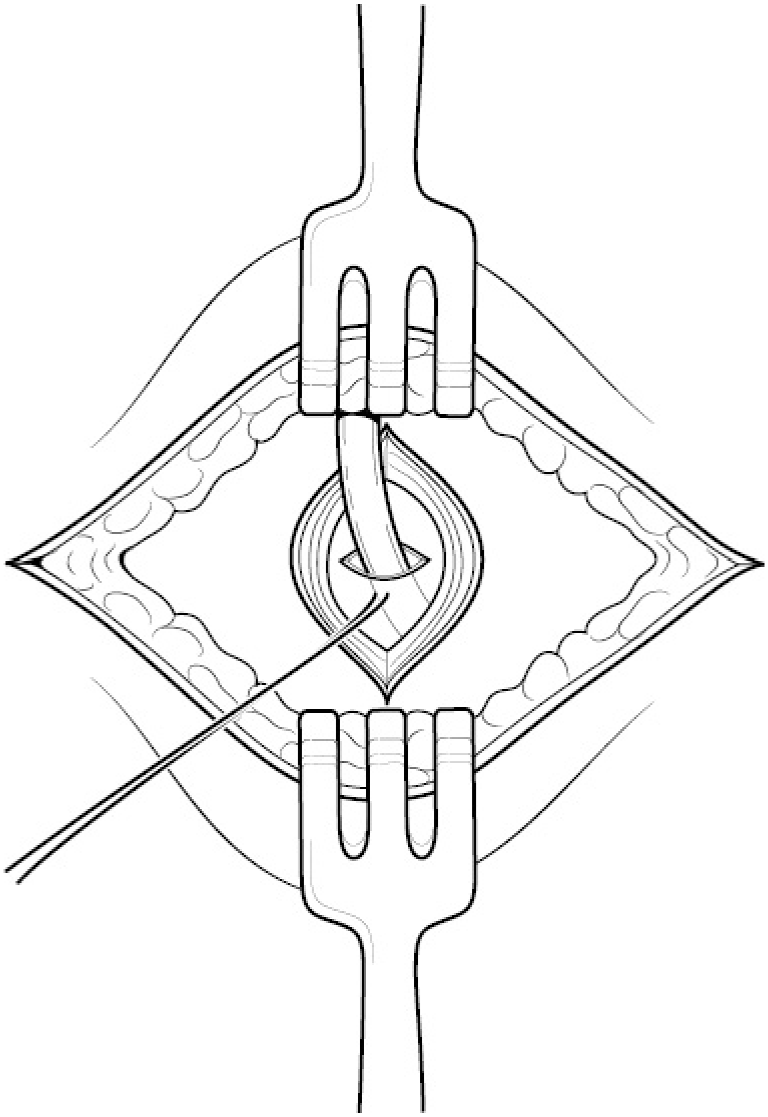
18.7

The shunt system is composed of two segments of ventriculoperitoneal shunt tubing connected together by a one-way pumping chamber. The system is primed with saline. Special attention is paid to orienting the pumping chamber toward the abdomen. The pleural end of the catheter is trimmed to the appropriate length and tunneled into the pleural space as far posterior as possible to allow for optimal dependent drainage. Care must be taken not to kink the catheter as it enters the pleural space. A long atraumatic clamp is then passed from the lower incision to the upper incision through the subcutaneous pocket (**Figure 18.8**). In small infants a low-profile ventriculoperitoneal shunt device works well. As the device may require active pumping, the pump/reservoir chamber should be placed over the thoracic cage to allow for ease of pumping.



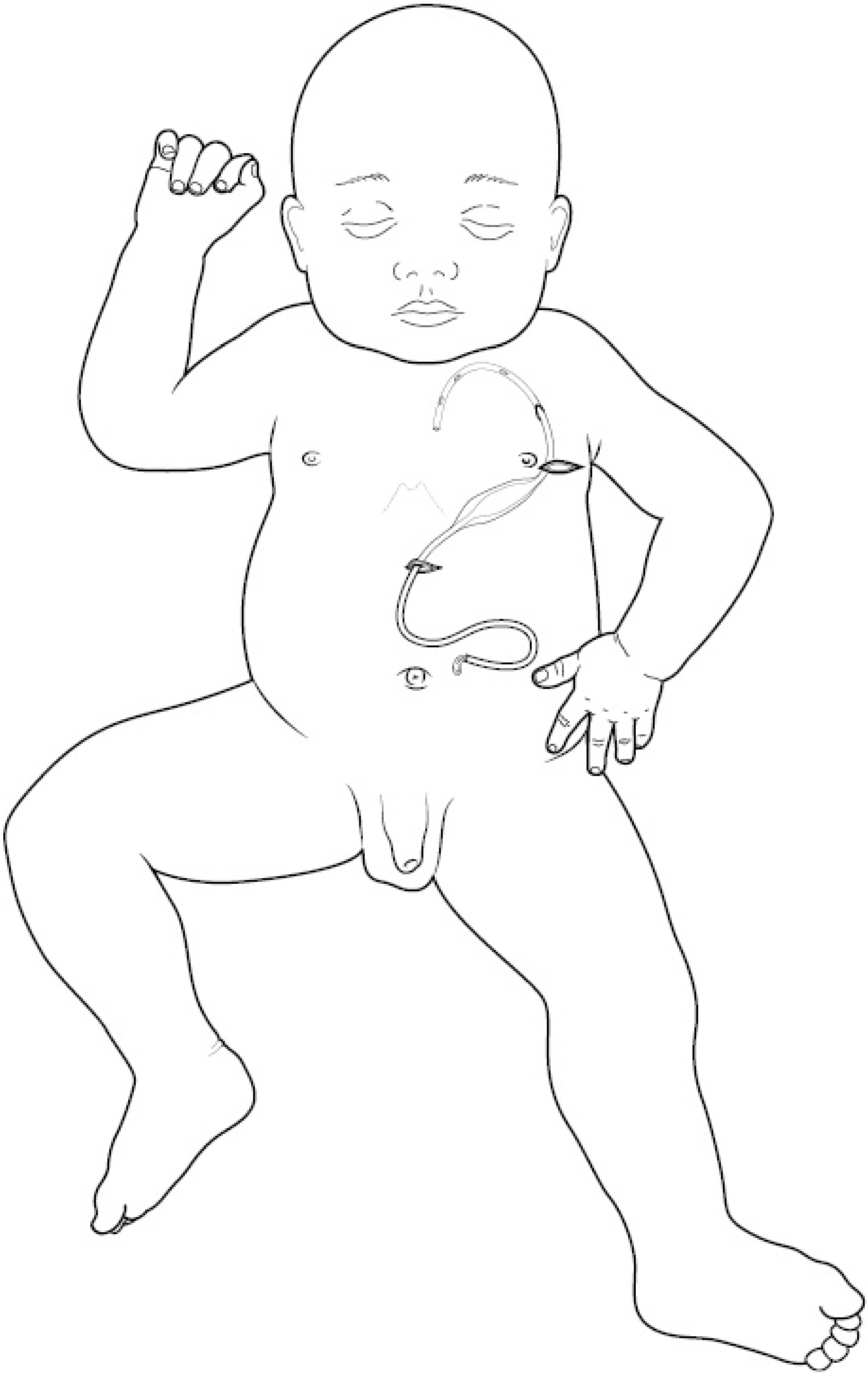
18.8

For the internal reservoir/chamber system, the peritoneal catheter and pumping chamber are drawn into the pocket by traction (**Figure 18.9**).



18.9

For the external chamber system, an additional stab incision is made near the xiphoid to create a long subcutaneous tunnel for the catheter prior to entry into abdominal cavity. The catheter entry and exit sites are secured to the skin with non-absorbable sutures. The peritoneal catheter is trimmed to the appropriate length and inserted into the abdominal cavity. The purse-string sutures are tied securely. The incisions are closed in layers using fine absorbable sutures (**Figure 18.10**).



18.10

POSTOPERATIVE CARE

The chamber should be manually pumped five to ten times every 8 hours to ensure catheter patency and removal of fluid. Over the ensuing weeks, the pumping sessions can be weaned accordingly. Pleuroperitoneal shunts can remain in place for months, if necessary, until pleural drainage ceases.

OUTCOME

The majority of chylothoraces, particularly those that are congenital in etiology, cease spontaneously under proper medical management. An operative strategy that includes thoracic duct ligation, often in conjunction with pleurectomy and pleurodesis, is effective in most of the remaining medically refractory cases. Chest tubes can be safely removed after the initiation of regular feeds with minimal output.

FURTHER READING

Beghetti M, La Scala G, Belli D, et al. Etiology and management of pediatric chylothorax. *J Pediatr* 2000; 136: 653–8.

Church JT, Antunez AG, Dean A, et al. Evidence-based management of chylothorax in infants. *J Pediatr Surg* 2017; 52: 907–12.

Gonzalez R, Bryner BS, Teitelbaum DH, et al. Chylothorax after congenital diaphragmatic hernia repair. *J Pediatr Surg* 2009; 44: 1181–5.

Helin RD, Angeles ST, Bhat R. Octreotide therapy for chylothorax in infants and children: A brief review. *Pediatr Crit Care Med* 2006; 7: 576–9.

Itkin M, Krishnamurthy G, Naim MY, et al. Percutaneous thoracic duct embolization as a treatment for intrathoracic chyle leaks in infants. *Pediatrics* 2011; 128: e237–41.

Katanyuwong P, Dearani J, Driscoll D. The role of pleurodesis in the management of chylous pleural effusion after surgery for congenital heart disease. *Pediatr Cardiol* 2009; 30: 1112–16.

Murphy MC, Newman BM, Rodgers BM. Pleuroperitoneal shunts in the management of persistent chylothorax. *Ann Thorac Surg* 1989; 48: 195–200.

Slater BJ, Rothenberg SS. Thoracoscopic thoracic duct ligation for congenital and acquired disease. *J Laparoendosc Adv Surg Tech A* 2015; 25(7): 605–7.

Teitelbaum DH, Teich S, Hirschl RB. Successful management of a chylothorax in infancy using a pleurectomy. *Pediatr Surg Int* 1996; 11: 166–8.

19

Mediastinal masses

BRENT R. WEIL AND ROBERT C. SHAMBERGER

PRINCIPLES AND JUSTIFICATION

Mass lesions of the mediastinum have multiple origins and may appear at any age throughout infancy, childhood, and adolescence. The mass may be cystic or solid, and of either congenital or neoplastic origin. The symptoms produced by a mediastinal mass are almost as diverse as the underlying pathology of these lesions, but most symptoms are due to the "mass effect" of the lesion which may compress the airway, vasculature, esophagus, or lung. Occasionally, they present with pain resulting from inflammation produced by infection or perforation of a cyst. Invasion of the chest wall by a malignant tumor will also produce pain. Many mediastinal lesions, in fact, are found as a radiographic abnormality on a study obtained for symptoms unrelated to the mass. Respiratory symptoms of expiratory stridor, cough, dyspnea, or tachypnea require urgent investigation. Cystic or solid lesions located at the carina may produce major airway obstruction. Lesions at this site are often "hidden" in the normal mediastinal shadow and may not be apparent on the anterior–posterior or lateral chest radiographs. Orthopnea and venous engorgement from superior vena caval syndrome occur with extensive involvement of the anterior mediastinum and are harbingers for respiratory obstruction upon induction of a general anesthetic. Less frequently, dysphagia from pressure on the esophagus is the presenting symptom. Neurologic symptoms from spinal cord compression or Horner's syndrome may occur with neurogenic tumors arising in the posterior mediastinum.

INDICATIONS FOR RESECTION

Management of mediastinal masses is determined by the presumed diagnosis. Cystic lesions in the anterior mediastinum are generally resected. Acute enlargement in thymic cysts has been noted following viral respiratory illnesses. Teratomas, because of their possible malignant degeneration, are also resected. Lymphatic malformations may secondarily involve the mediastinum with their predominant component in the cervical–facial area. Isolated mediastinal involvement is seen in less than 5% of cases. Pericardial cysts are the most innocent of these lesions and, if well demonstrated on scans and radiographs, often are simply followed because they rarely increase in size and are unlikely to compress any vital structures.

The solid lesions require establishment of a histopathologic diagnosis. The most common solid tumor in the anterior mediastinum is Hodgkin lymphoma followed by non-Hodgkin lymphoma. Primary treatment of these tumors is by chemotherapy often in conjunction with radiotherapy; the surgeon's role is to establish the diagnosis. The primary treatment of the malignant germ cell tumors of the mediastinum is also chemotherapy, with surgical resection of residual masses after treatment. Teratomas or dermoids are the only neoplastic lesions which require primary resection as they may become secondarily infected or undergo malignant degeneration if they contain immature elements. Retrosternal thyroid goiters are generally resected through the neck. These latter tumors and substernal extension of cervical lymphatic malformations (cystic hygromas) can often be quite readily removed from a suprasternal incision; with progressive retraction and dissection, one can remove quite sizable retrosternal masses originating in the neck.

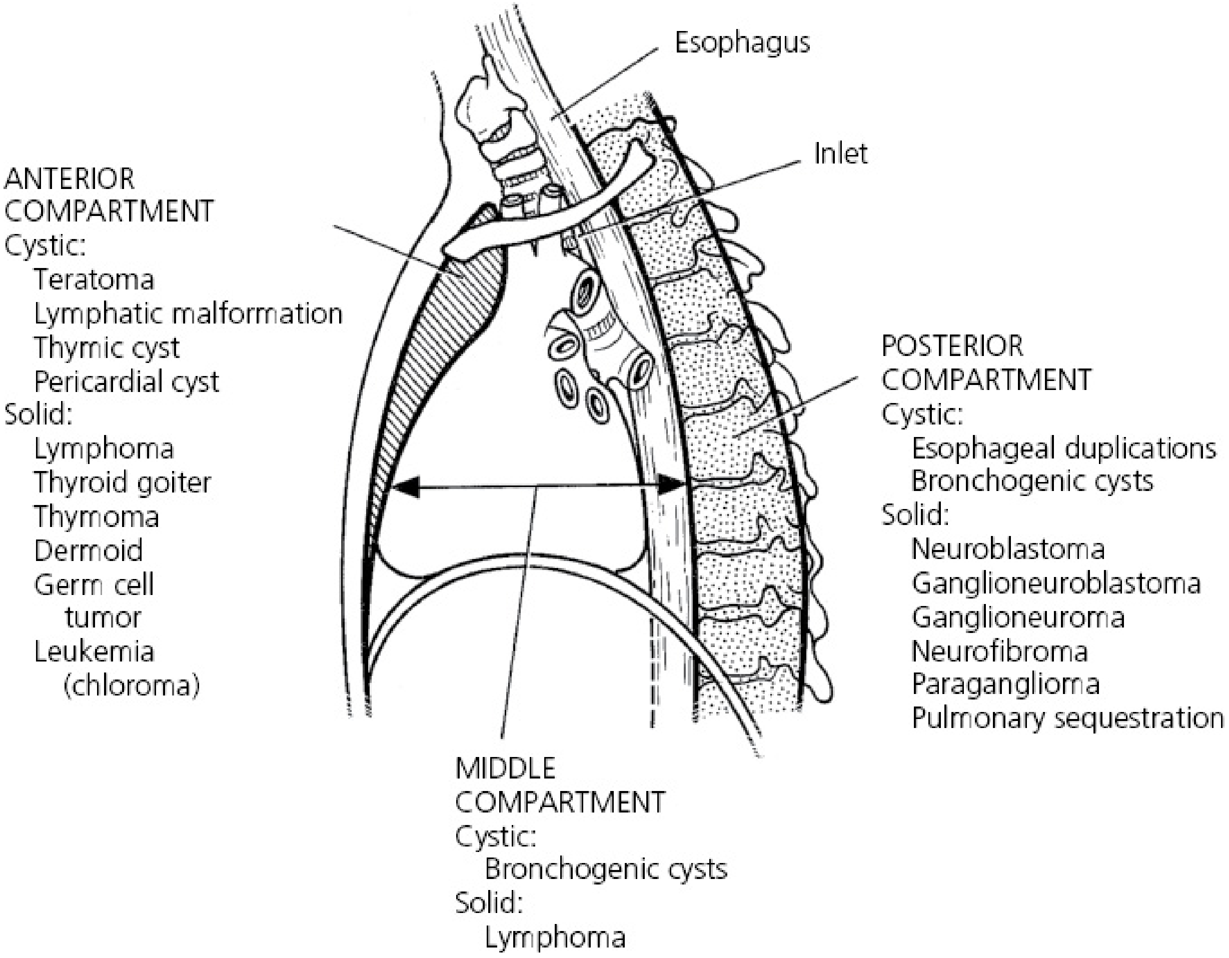
Bronchogenic cysts and esophageal duplications arise in the middle and posterior mediastinum. They develop in the embryo during separation of the aerodigestive systems. Bronchogenic cysts are generally lined by respiratory epithelium and esophageal duplications by intestinal mucosa, but ectopic mucosa may be present in both lesions. These should be resected because of their potential for growth with accumulation of secretions. They can also become secondarily infected or develop malignancy. Lesions with gastric mucosa can erode into the bronchus, esophagus, or pleural cavity.

Solid tumors in the posterior mediastinum should be resected. For thoracic neuroblastoma, this is a major component of its treatment. The requirement for radiation therapy or chemotherapy will depend on the age of the child, the presence of metastatic disease, and the cytogenetic findings of the tumor, particularly amplification of the *MYCN* oncogene or normal ploidy, both of which predict an aggressive tumor. Ganglioneuroma and ganglioneuroblastoma, while benign tumors, may grow locally, may erode the ribs, and may extend into the spinal canal producing neurologic symptoms. While these benign lesions are often identified when asymptomatic, resection generally is recommended to establish diagnosis and prevent local extension. A paraganglioma (extra-adrenal pheochromocytoma) should be removed to control the systemic manifestations of neuropeptide production. The patient should be well prepared for surgery with alpha- and beta-blocking agents and volume repletion. Pulmonary sequestrations are generally resected to obtain a definitive pathologic diagnosis, to treat the arteriovenous shunt which may be present, and to avoid infection in the intralobar sequestrations.

DIAGNOSIS

The preoperative diagnosis of a mediastinal mass can be established quickly with only a few diagnostic studies. Anterior–posterior and lateral chest radiographs will demonstrate the area of the mediastinum in which the mass arises. The location and knowledge of whether the mass is cystic or solid and the age of the patient will often allow an accurate diagnosis to be made.

Lesions occurring in each of the three compartments of the mediastinum are shown in **Figure 19.1**, grouped by their cystic or solid nature. Further investigation is determined by the location of the lesion.


Esophagus
Inlet
ANTERIOR COMPARTMENT
Cystic:
Teratoma
Lymphatic malformation
Thymic cyst
Pericardial cyst
Solid:
Lymphoma
Thyroid goiter
Thymoma
Dermoid
Germ cell tumor
Leukemia (chloroma)
POSTERIOR COMPARTMENT
Cystic:
Esophageal duplications
Bronchogenic cysts
Solid:
Neuroblastoma
Ganglioneuroblastoma
Ganglioneuroma
Neurofibroma
Paraganglioma
Pulmonary sequestration
MIDDLE COMPARTMENT
Cystic:
Bronchogenic cysts
Solid:
Lymphoma

19.1

Masses in the anterior compartment

Masses in the anterior mediastinum may produce respiratory symptoms and cause compression of the trachea. Computed tomography (CT) scan is generally best for evaluation of masses in this area; it defines the cystic or solid nature of the lesion and most accurately demonstrates the extent of tracheal compression. The extent to which the trachea is compressed will determine the safety of anesthesia required for further diagnosis or resection.

Cystic lesions may be differentiated by their structure and location. A teratoma generally has both cystic and solid components with areas of varying density that are well demonstrated on the CT scan. Most lymphatic malformations have multiple cysts with very thin walls which often extend up into the neck. Only a small proportion is limited entirely to the anterior mediastinum. Thymic cysts are often single, thin-walled, and contiguous with the thymus. Pericardial cysts arise in the inferior portion of the chest adjacent to the pericardium.

Solid anterior mediastinal lesions are also easily assessed on a CT scan. A dermoid or entirely solid teratoma has areas of varying fat and water density and often calcification. A substernal thyroid goiter arises from the thyroid gland and extends into the retrosternal space. Thymomas are extremely rare in children. Lymphomas involve multiple nodal sites. The CT scan also defines lymph node enlargement in the pulmonary hilum and pulmonary parenchymal lesions. Germ cell tumors are uncommon, arising primarily in teenagers or young adults. They are usually diagnosed with serum markers and can be further assessed with CT or magnetic resonance imaging (MRI). These tumors can occasionally arise from within the pericardium, often at the root of the aorta.

Masses in the middle compartment

A bronchogenic cyst at the carina may be "hidden" in the mediastinal shadow on the chest radiograph despite the lesion being large enough to produce significant respiratory distress. Fluoroscopy of the infant will demonstrate compression and anterior displacement of the airway, and ingestion of barium into the esophagus will demonstrate posterior displacement of the esophagus and confirm the presence of a space-occupying lesion.

Masses in the posterior compartment

The main cystic lesions in this area are bronchogenic cysts and esophageal duplications which are typically ovoid in shape and may be suspected on routine radiographs.

The solid neural tumors have a fusiform shape and are based in the posterior sulcus between the vertebral bodies and the ribs. The age of the patient will give some hint of the diagnosis: Neuroblastomas and ganglioneuroblastomas arise more often in infants. Ganglioneuromas occur in older children and are generally asymptomatic, but they can extend into the spinal canal and produce neurologic symptoms. Neurofibromas arise primarily in conjunction with neurofibromatosis (von Recklinghausen's disease) and are often associated with scoliosis. Paragangliomas may arise in the posterior mediastinum, although they are rare. They often present with symptoms related to catecholamine secretion, particularly paroxysmal hypertension, diaphoresis, and palpitations. Extralobar pulmonary sequestrations arise in the posterior mediastinum with arterial supply from the aorta. They generally can be distinguished by a triangular shape.

MRI is often used to evaluate patients with masses in the posterior mediastinum, because it provides a better definition of possible extension of the tumor into the spinal canal than a CT scan. It is important to identify this extension prior to surgical resection. In infants and young children in whom neuroblastoma is a major diagnostic concern, evaluation for metastatic disease is also important. This should include a bone marrow biopsy, metaiodobenzylguanidine (MIBG) scan, and urine should be collected to measure catecholamines which are elevated in 95% of infants and children with neuroblastoma. For extensive posterior mediastinal masses, particularly left-sided lesions, it is reasonable to consider preoperative angiography (traditional or with cross-sectional imaging) to define the artery of Adamkiewicz, as injury to this vessel has been implicated as a cause of postoperative spinal ischemia in rare instances. The majority of esophageal and bronchogenic cysts are not associated with vertebral anomalies, but some rare cases in which large duplication cysts originate from the stomach, pancreas, or duodenum and extend into the thoracic cavity will demonstrate abnormalities of the vertebrae.

PREOPERATIVE

Solid lesions require further histopathologic diagnosis. The most common solid tumor in the anterior mediastinum is Hodgkin lymphoma followed by non-Hodgkin lymphoma. Other areas of lymph node involvement besides the mediastinum, particularly the neck, should be sought where biopsy could be more easily performed. In those rare instances where only the mediastinum is involved, a germ cell tumor should be suspected and serum alpha-fetoprotein (AFP) and human chorionic gonadotrophin (hCG) levels should be obtained. In cases where there is no extrathoracic tumor, either a needle biopsy with radiographic guidance or a limited anterior thoracotomy or thoracoscopy may be required to establish a tissue diagnosis. The rare chloroma of leukemia presenting as a mediastinal mass can be diagnosed with the initial complete blood count and bone marrow biopsy. Serum metanephrine levels should be obtained for suspected paragangliomas. For suspected neuroblastomas, urine catecholamine measurement, MIBG scan, and/or bone marrow biopsy should be considered.

Preparation for surgery

The child should be prepared for surgery after completion of diagnostic studies. If a bronchogenic cyst is compressing the airway sufficiently to produce pneumonia or respiratory distress, no undue delay should occur. Appropriate antibiotic coverage and physiotherapy should be instituted for pneumonia. Preliminary bronchoscopy should be avoided in these patients, because a tenuous airway in an infant or child will be further compromised by manipulation. Catecholamine-secreting tumors, primarily paragangliomas, require institution of alpha- and beta-blocking agents and volume repletion. Direct involvement of the bronchus is very rare and compression of the airway can be defined most safely radiographically. In the occasional case of a thymoma and associated myasthenia gravis, the neuromuscular deficit should be minimized as much as possible prior to surgical intervention.

Anesthesia

Anesthesia is of major concern primarily for solid lesions of the anterior compartment which often compress the airway. A cross-sectional tracheal area of less than 50% of that expected for age or a peak expiratory flow rate of less than 50% of predicted suggests that a child is at significant risk for respiratory and or cardiovascular collapse upon induction of anesthesia. Children with either of these two findings must be limited to local anesthesia with sedation; general anesthesia (particularly with paralytic agents) must be avoided at all costs. Bronchogenic cysts in the area of the carina may also cause significant airway obstruction in infants, but the endotracheal tube can generally be passed down one of the mainstem bronchi to provide adequate ventilation until the pressure is relieved. This maneuver may not be feasible in children with a solid mass compressing the airway.

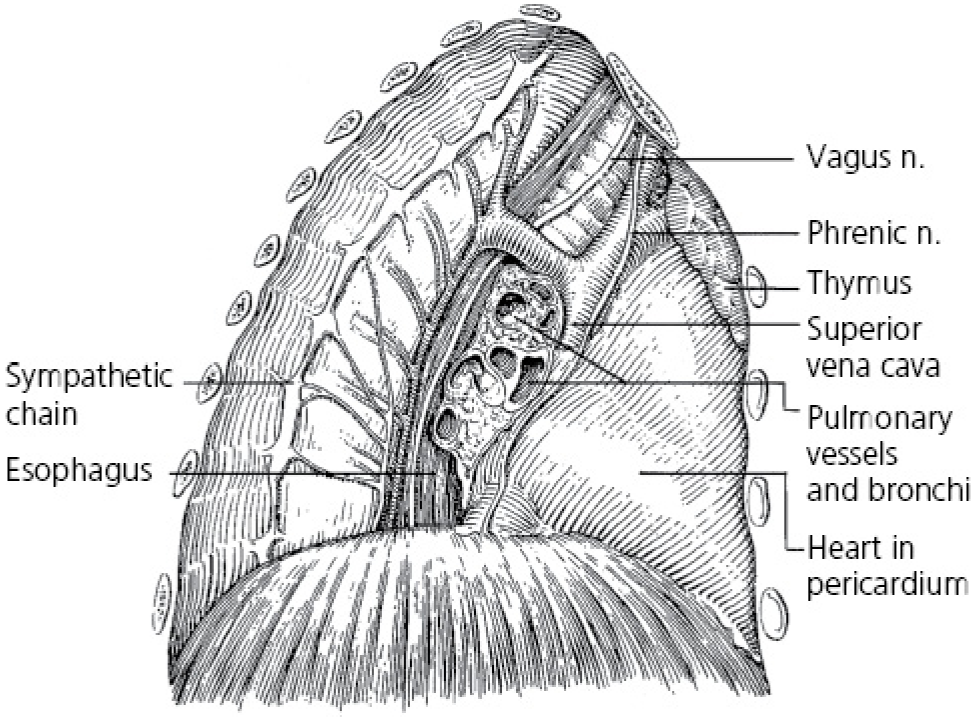
Appropriate monitoring of these patients requires transcutaneous oximetry and, in those children requiring extensive resections, a central venous line and arterial pressure monitoring are utilized. Uncuffed endotracheal tubes are routinely used in younger children to avoid any injury to the airway from pressure. An "air leak" should be present around the endotracheal tube to confirm that pressure on the subglottic mucosa, the narrowest segment of an infant's upper airway, is not excessive.

For thoracoscopic procedures, single-lung ventilation will often facilitate the resection. Double-lumen endotracheal tubes are available down to 26 Fr caliber and may be used on children of over 25 kg. In smaller children or infants, mainstem intubation of the contralateral bronchus or placement of a balloon catheter (bronchial blocker) on the ipsilateral side will allow deflation of the lung to facilitate exposure.

Choice of approach and applied anatomy

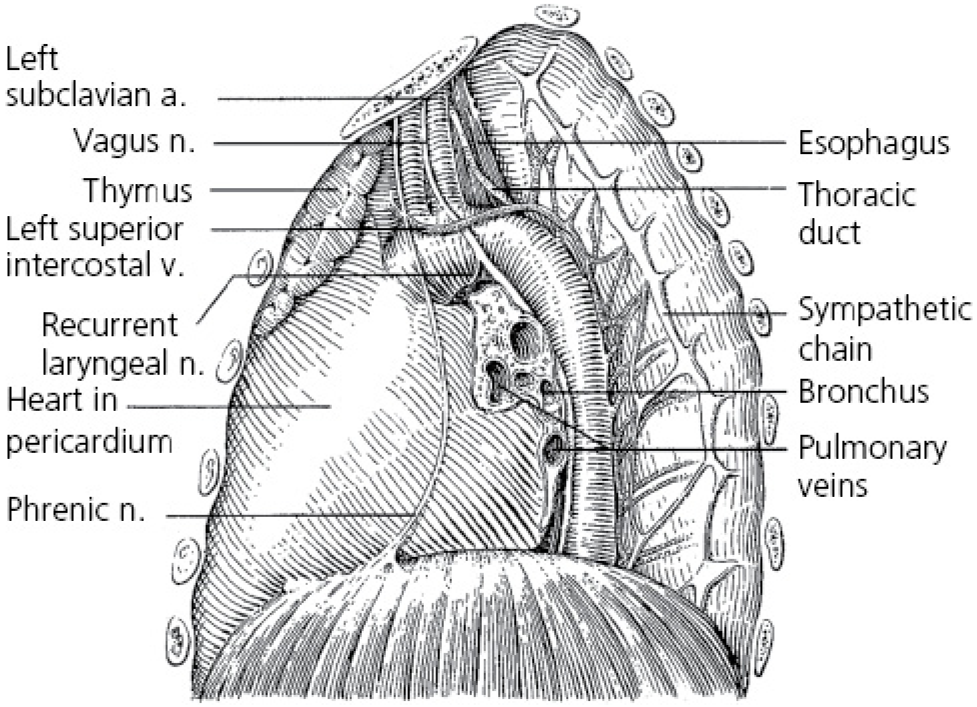
The approaches to these masses are based primarily on their location and nature; most may be resected through a posterolateral thoracotomy or thoracoscopically. If a teratoma or a dermoid is primarily located in the midline, it may be most easily resected through a median sternotomy. These lesions, however, are often asymmetric and prolapse into one of the hemithoraxes allowing them to be resected from that side. This is also true of a thymic cyst. Extensive lymphatic malformations, if they extend into the thoracic cavity, are also best dealt with through a thoracotomy or thoracoscopically in instances when surgical management becomes necessary. Sternotomy should be avoided for suspected lymphomas because compression of the airway can occur when the sternum is closed after biopsy of the mass. Posterolateral thoracotomy and thoracoscopic approaches are appropriate for lesions of the posterior compartment depending on the size of the lesion and experience of the surgeon. When patients exhibit neurologic symptoms and/or spinal cord compression, extension into the spinal canal from benign tumors may require a preliminary laminectomy with resection of the tumor, or a combined laminectomy and thoracotomy as initial resection of the thoracic component has been reported to result in swelling of the residual segment within the spinal canal and associated neurologic symptoms. In asymptomatic children with confirmed or suspected ganglioneuroma, ganglioneuroblastoma, or neuroblastoma, it is reasonable to leave behind small amounts of foraminal tumor to avoid unnecessary surgical morbidity. The anatomic considerations and steps of the procedures are identical for open or thoracoscopic resections.

The major structures of concern on the right side of the mediastinum are shown in **Figure 19.2**. Particular care should be taken to preserve the phrenic nerve, avoiding loss of diaphragmatic function. The upper mediastinum and carina are most readily approached from the right side because of the aortic arch and its branches on the left.


Vagus n.
Phrenic n.
Thymus
Superior vena cava
Pulmonary vessels and bronchi
Heart in pericardium
Sympathetic chain
Esophagus

19.2

On the left side, in addition to the phrenic nerve, attention must be paid in the upper mediastinum to the course of the vagus nerve and the recurrent laryngeal nerve, which loops around the aortic arch before it ascends to the larynx (**Figure 19.3**).


Left subclavian a.
Vagus n.
Thymus
Left superior intercostal v.
Recurrent laryngeal n.
Heart in pericardium
Phrenic n.
Esophagus
Thoracic duct
Sympathetic chain
Bronchus
Pulmonary veins

19.3

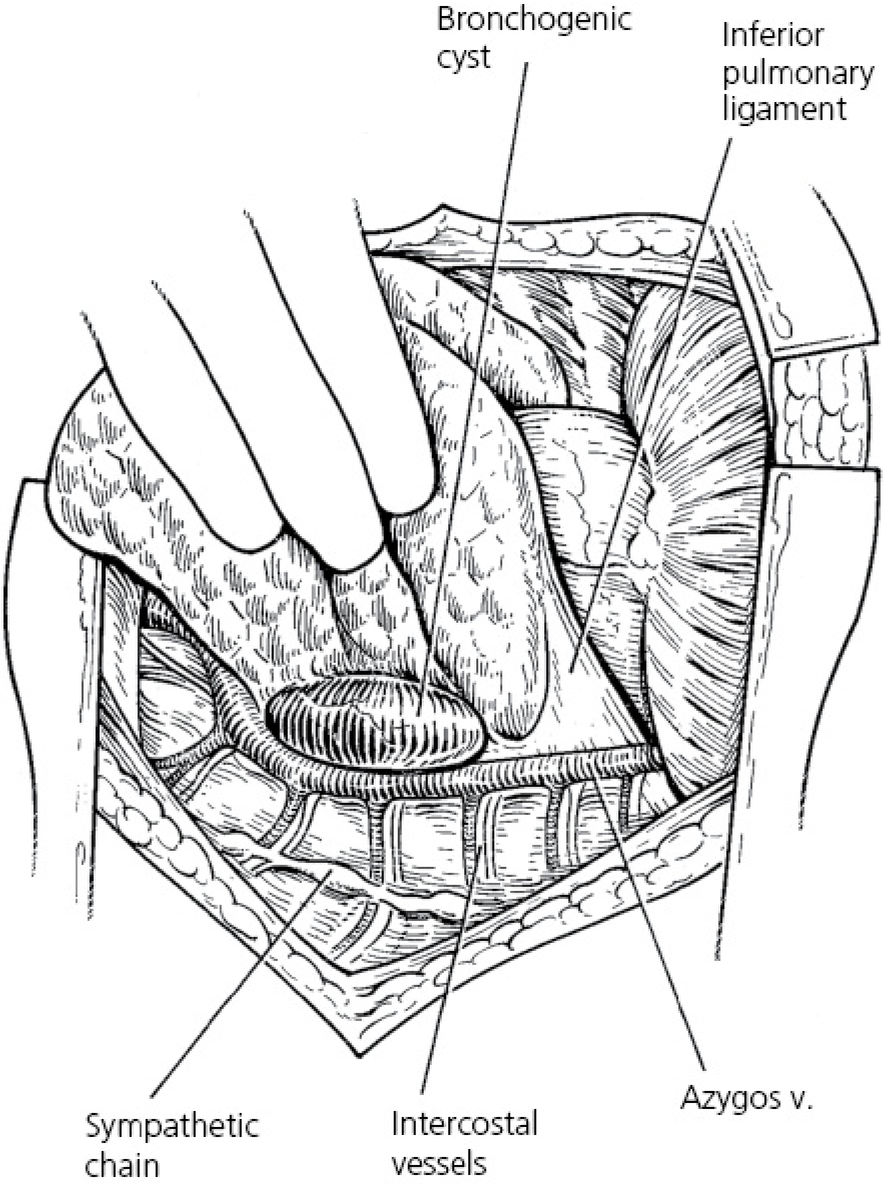
OPERATIONS

Posterolateral thoracotomy and thoracoscopy

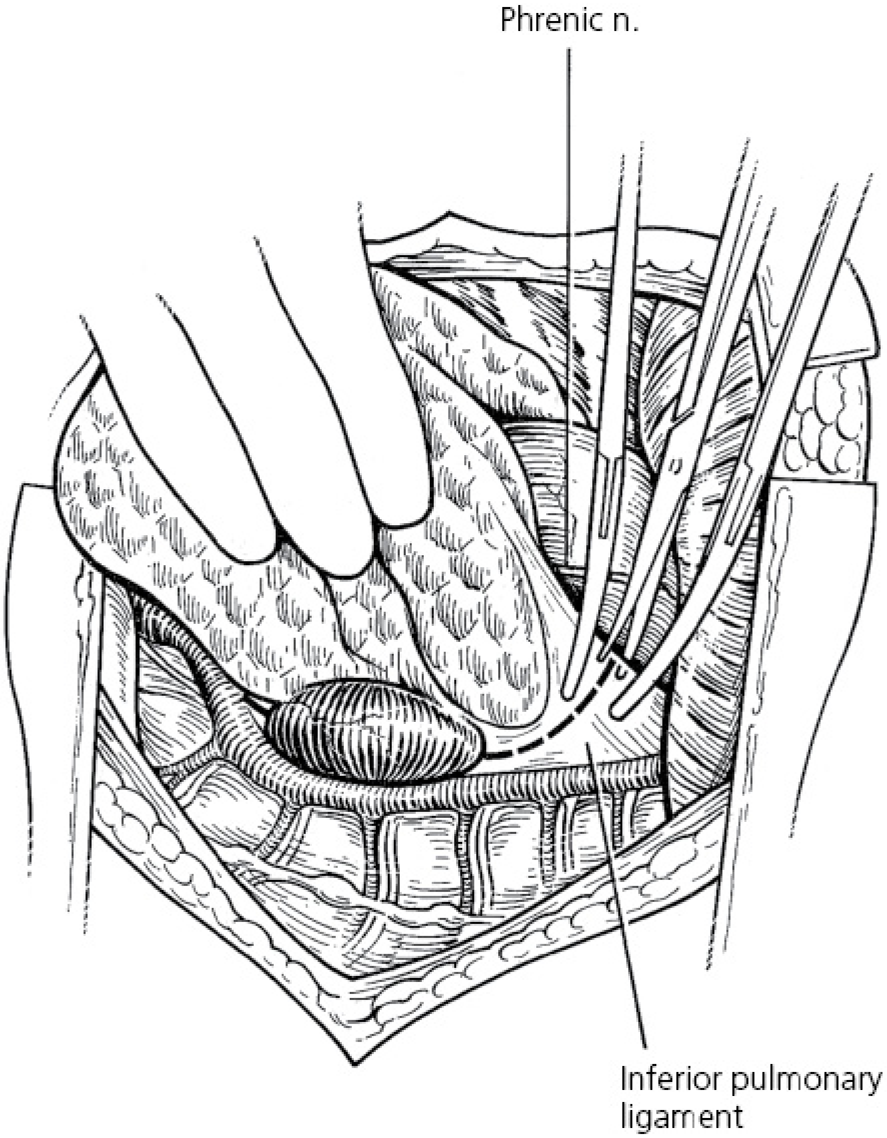
The technique of posterolateral thoracotomy is discussed in Chapter 16.

Bronchogenic cysts

The thoracic cavity is explored to identify the extent of the mass and its relationship with the vital intrathoracic structures. The lung is retracted anteriorly to expose the mass (**Figure 19.4**). During the resection of cystic lesions, aspiration is unnecessary unless the airway is compressed or the lesion is too large for safe dissection. Keeping the cyst filled with secretions often actually facilitates its dissection. These lesions can be resected by either open or thoracoscopic methods. The pleura around a cystic lesion is incised first (**Figure 19.5**).

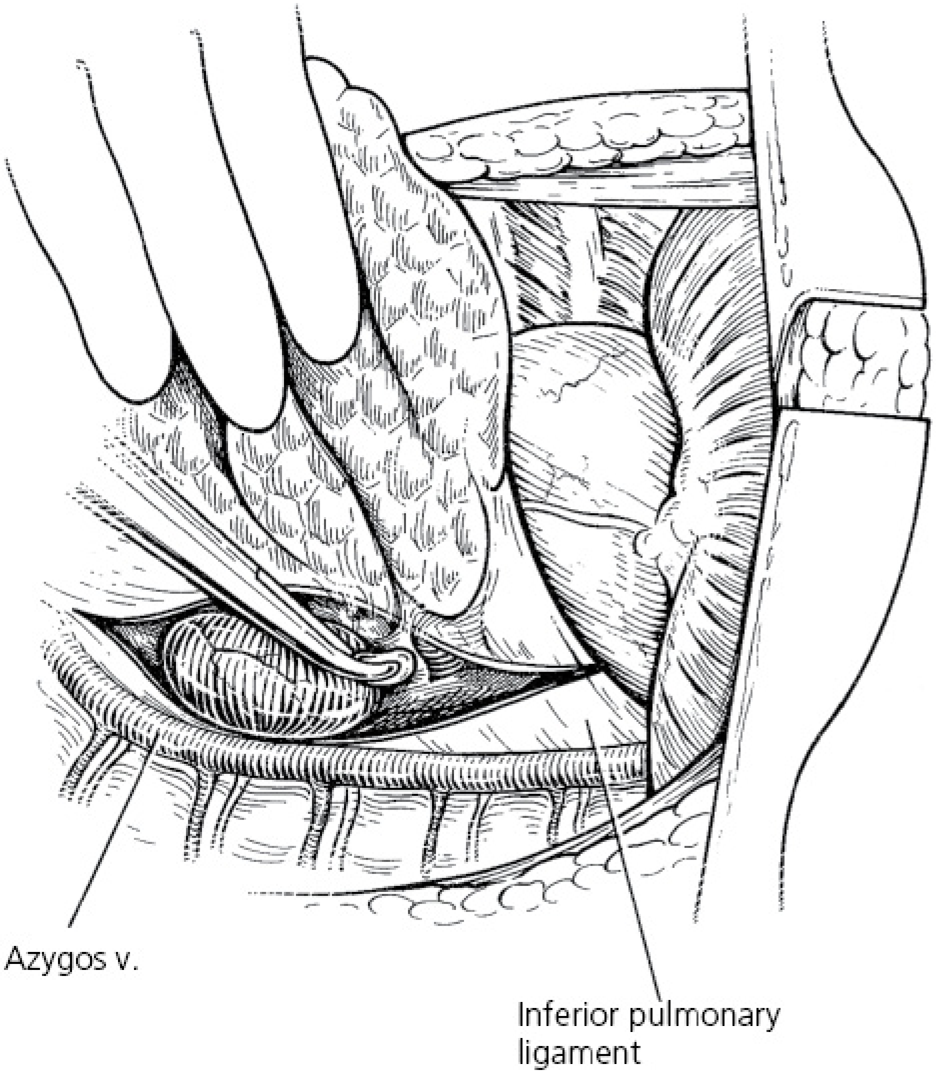

Bronchogenic cyst
Inferior pulmonary ligament
Sympathetic chain
Intercostal vessels
Azygos v.

19.4



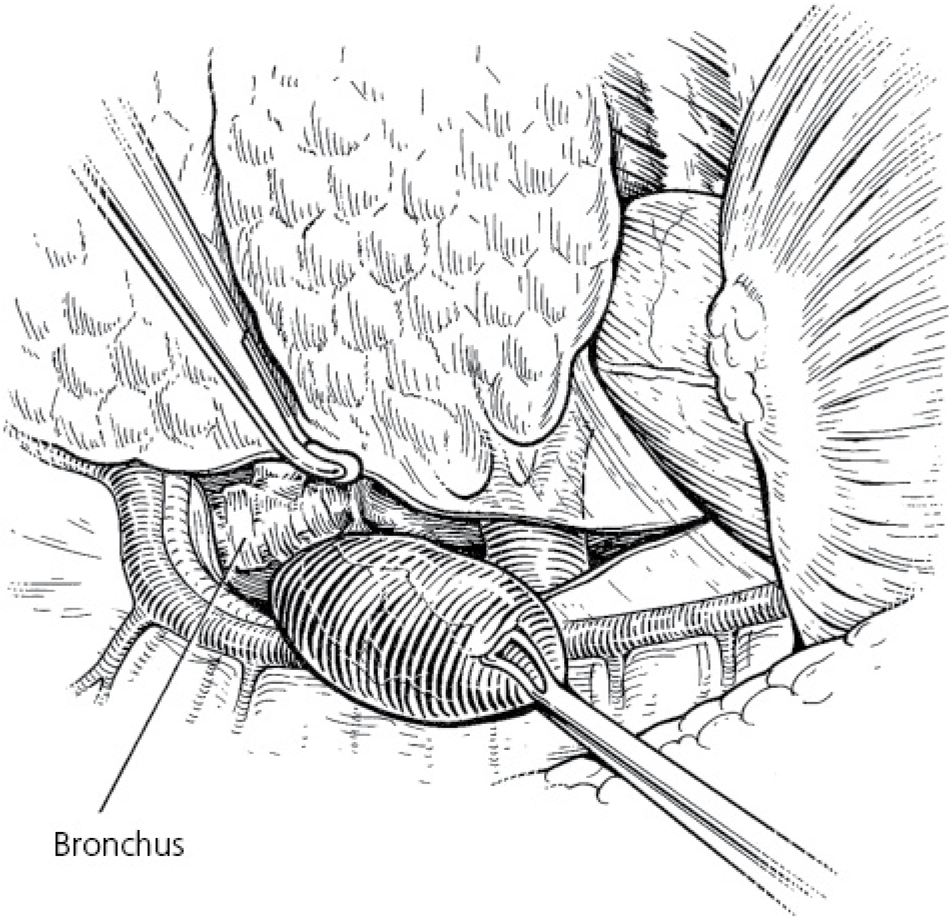
19.5

Bronchogenic cysts generally lie adjacent and posterior or lateral to the bronchus or trachea, but direct communication is extremely rare. They can be easily dissected away from surrounding structures and removed intact (**Figure 19.6**). Extreme caution must be taken to avoid injury to the membranous part of the trachea and bronchus from dissection or cautery. Delayed recognition of this injury has been reported after thoracoscopic resections.


Azygos v.
Inferior pulmonary ligament

19.6

Cysts associated with the right bronchus, carina, and central left bronchus are best approached through the right chest; only more peripheral lesions of the left bronchus lateral to the aortic arch are resected through the left chest (**Figure 19.7**).

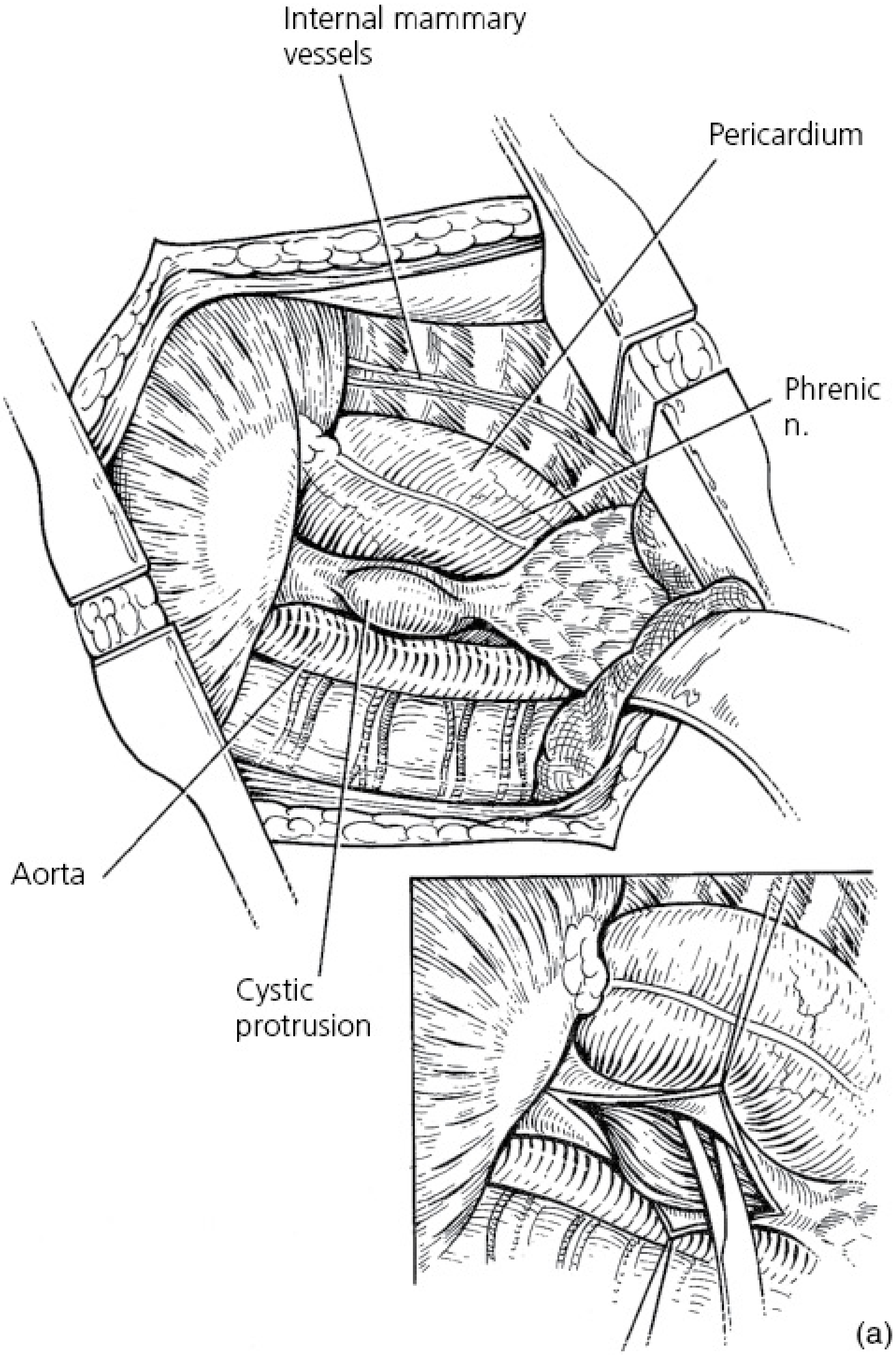


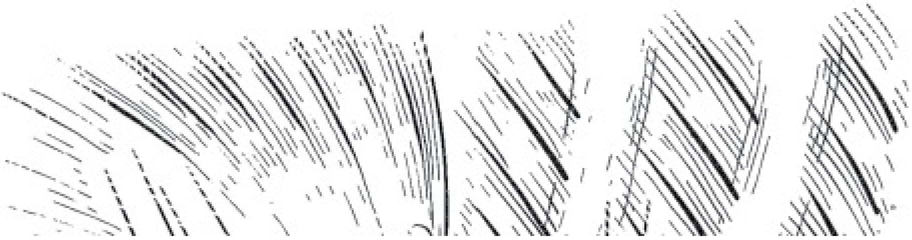
19.7

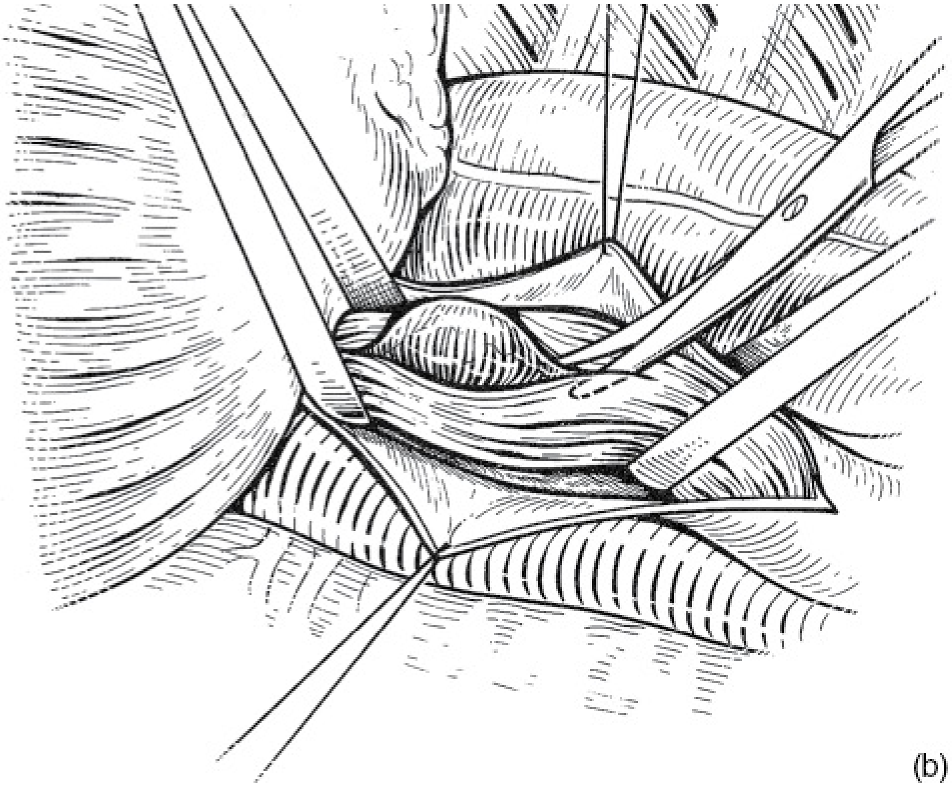
An inflammatory reaction around the cyst suggests ectopic gastric mucosa within the cyst or secondary infection of the cyst. Only rarely, when acid produced by the gastric mucosa has eroded through the cyst wall, will it be densely adherent to either the bronchus or esophagus. Significant hemorrhage or pulmonary reaction can occur in this situation. Cysts with gastric mucosa eroding into the bronchus or the esophagus may present with hemoptysis, hematemesis, or pain.

Esophageal duplications

Esophageal duplications are generally surrounded by esophageal muscle. They are best approached through the side of the thorax into which they protrude. These can also be resected by either open or thoracoscopic methods. The pleura is incised first over the esophagus and then the muscle overlying the cystic lesion is opened longitudinally (**Figure 19.8a and b**). It is particularly helpful to keep the cyst intact during this dissection.

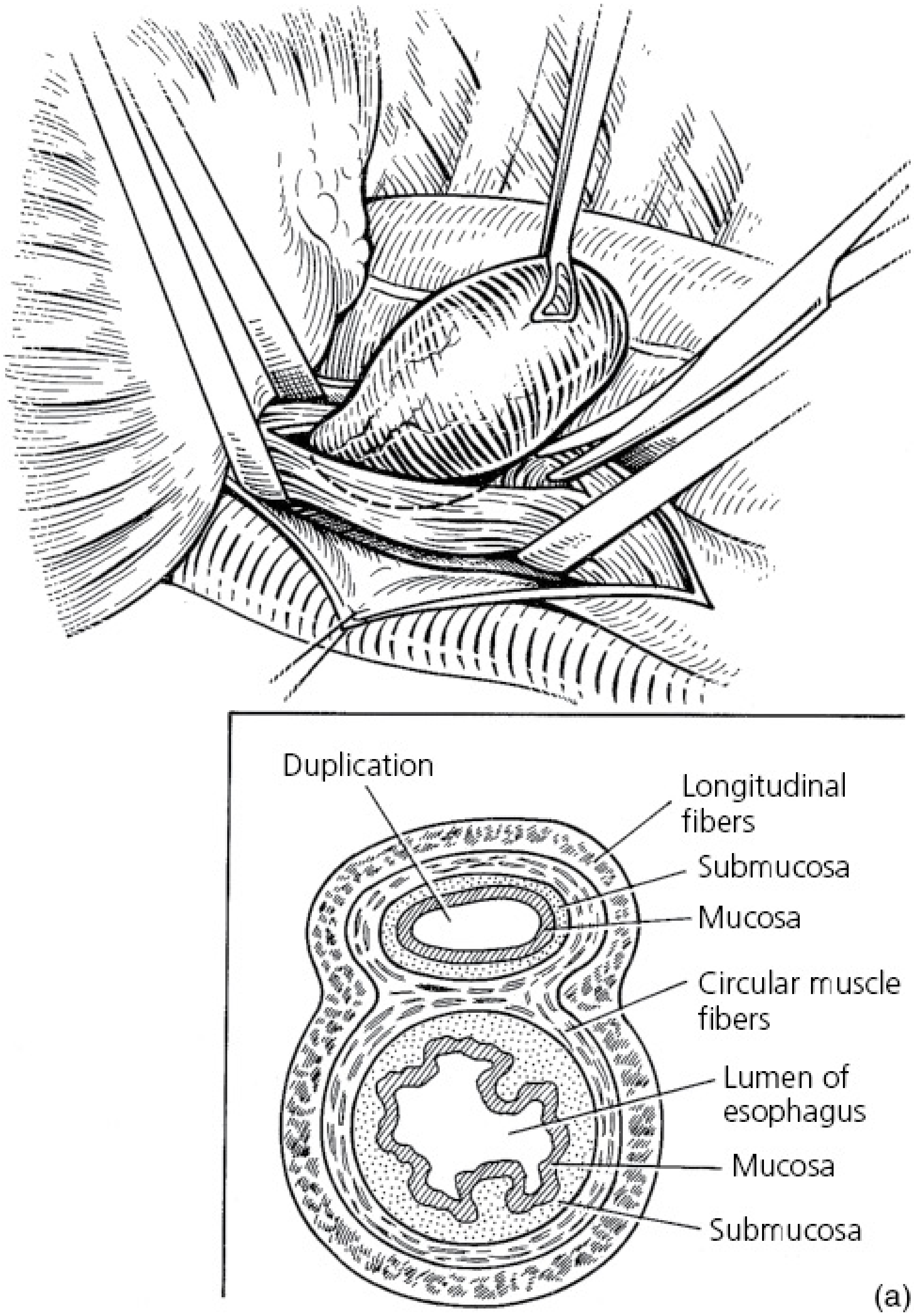

Internal mammary vessels
Pericardium
Phrenic n.
Aorta
Cystic protrusion
(a)



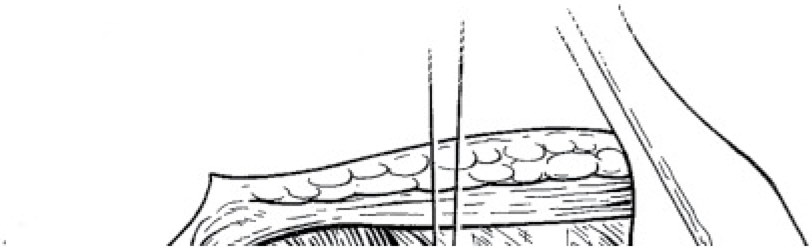


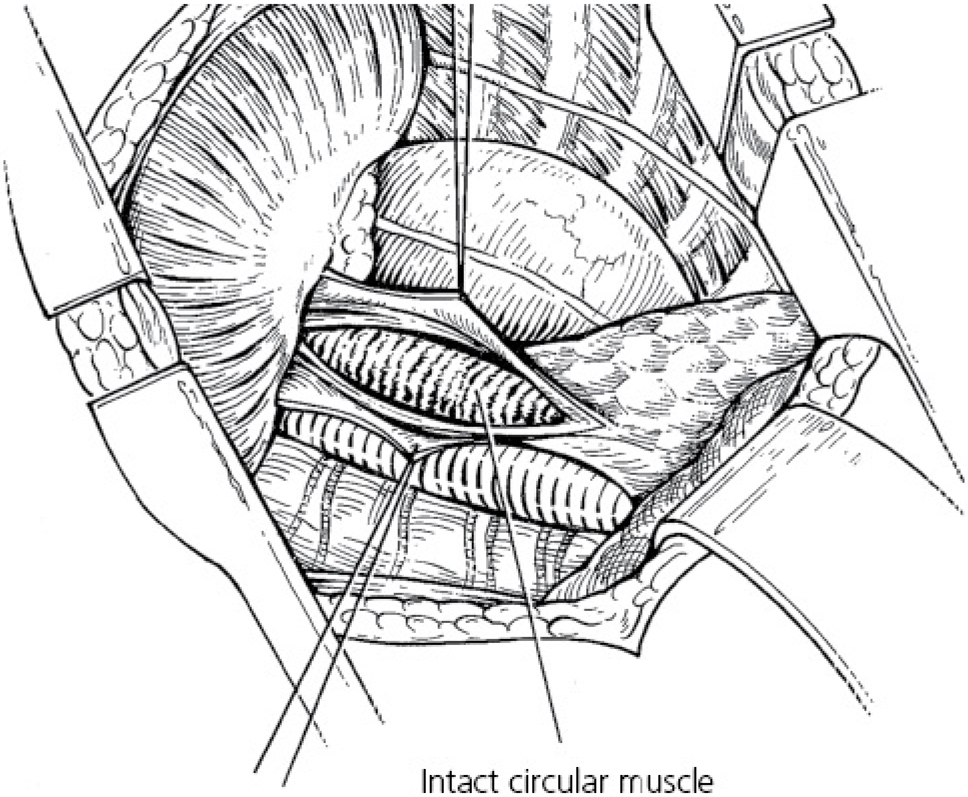
19.8a,b

Submucosal resection is the safest plane of dissection of the cyst from the esophagus to avoid an untoward entrance into the esophageal lumen. A single muscular layer comprises the common wall between the esophagus and the duplication (**Figure 19.9a**). After completion of the resection, a nasogastric tube or bougie can be passed to confirm that the mucosa is intact. Reapproximation of the muscular wall may prevent the formation of a diverticulum which occurs following resection of the cyst in some cases (**Figure 19.9b**).


Duplication
Longitudinal fibers
Submucosa
Mucosa
Circular muscle fibers
Lumen of esophagus
Mucosa
Submucosa

(a)



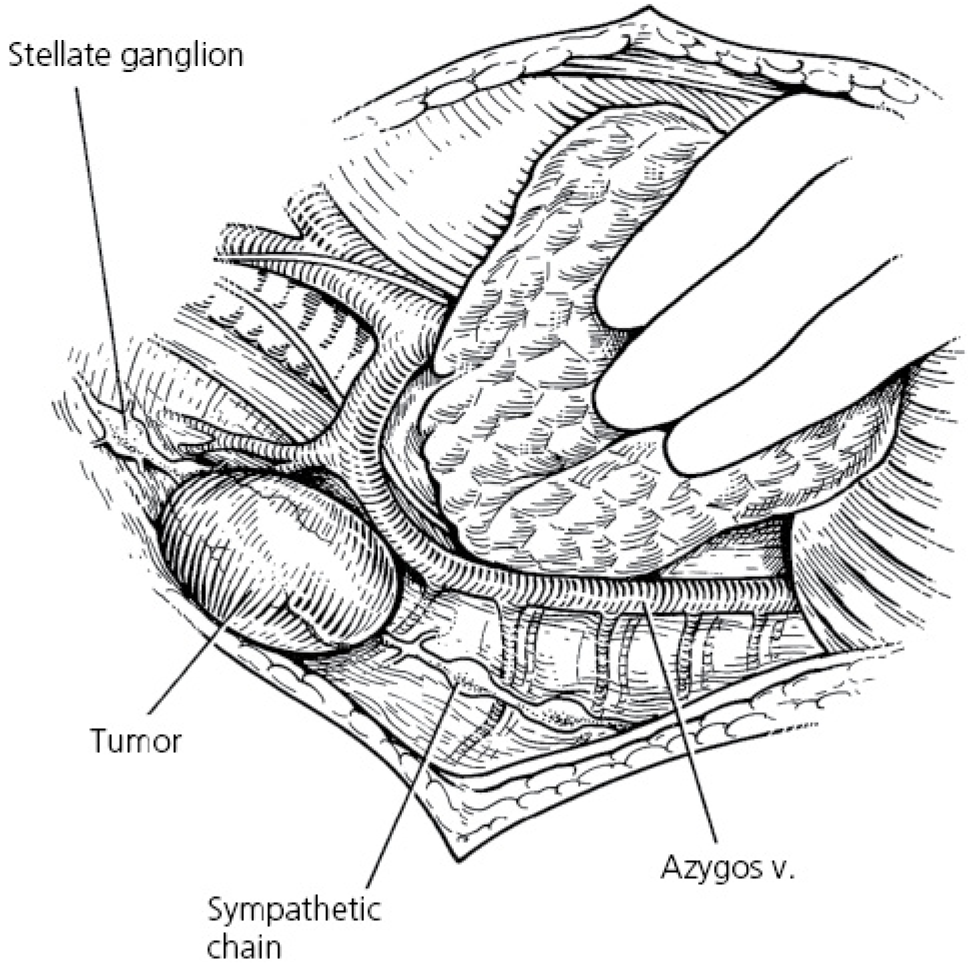


19.9a,b

Bronchogenic cysts and esophageal duplications should have their mucosa removed in its entirety. Any remaining mucosa may cause the cyst to recur. Aspiration or sclerosis of these lesions is not recommended because the mucosal surface will regenerate and produce a recurrent mass. The risk of development of malignancy in the mucosa must always be considered. In cases of a common wall with the posterior trachea or bronchus, it may be necessary to marsupialize a small portion to avoid injury to the trachea.

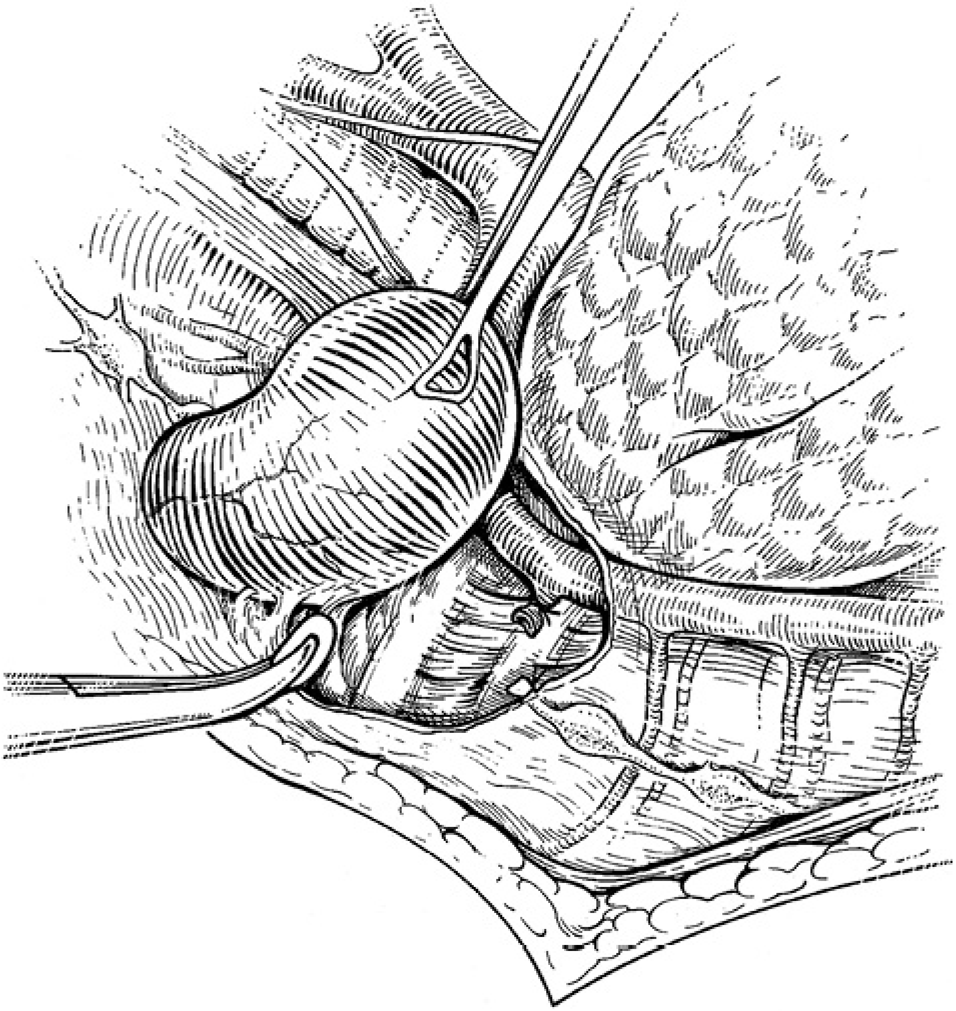
Solid posterior masses

Solid posterior mediastinal lesions are most frequently neurogenic in origin. They generally are broad-based and adhere to the ribs, intercostal muscles, and lateral surface of the vertebral bodies. They commonly arise from the sympathetic chain and may involve the stellate ganglion (**Figure 19.10**). Resection of this ganglion with an apical tumor will produce Horner's syndrome with apparent ptosis, miosis, and anhidrosis. The family and child should be forewarned of this possibility.



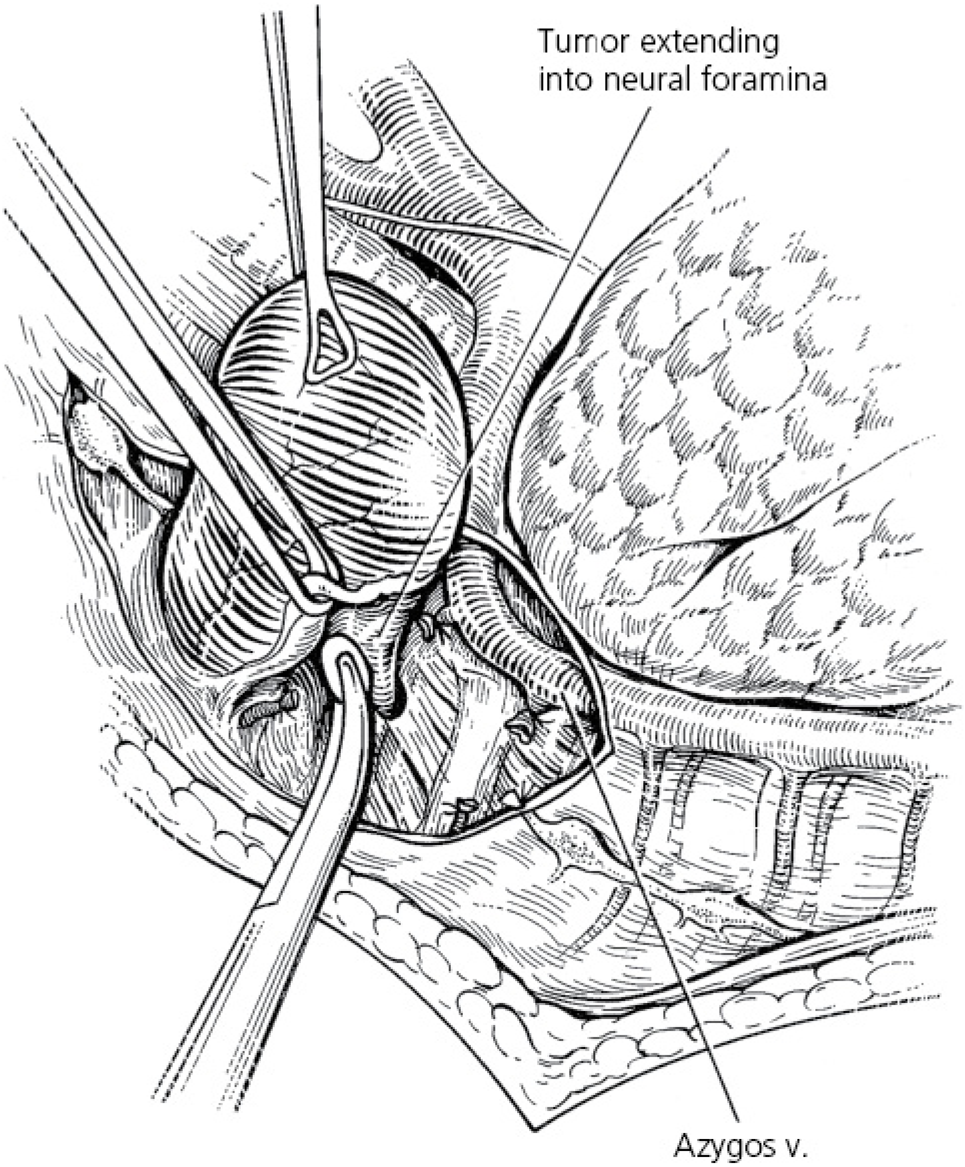
19.10

After incision of the pleura around the periphery, blunt dissection is used to elevate the tumor off the ribs (**Figure 19.11**). This will facilitate identification of the plane between the tumor and the intercostal muscles. These lesions may be densely adherent to the chest wall, making thoracoscopic resection challenging.



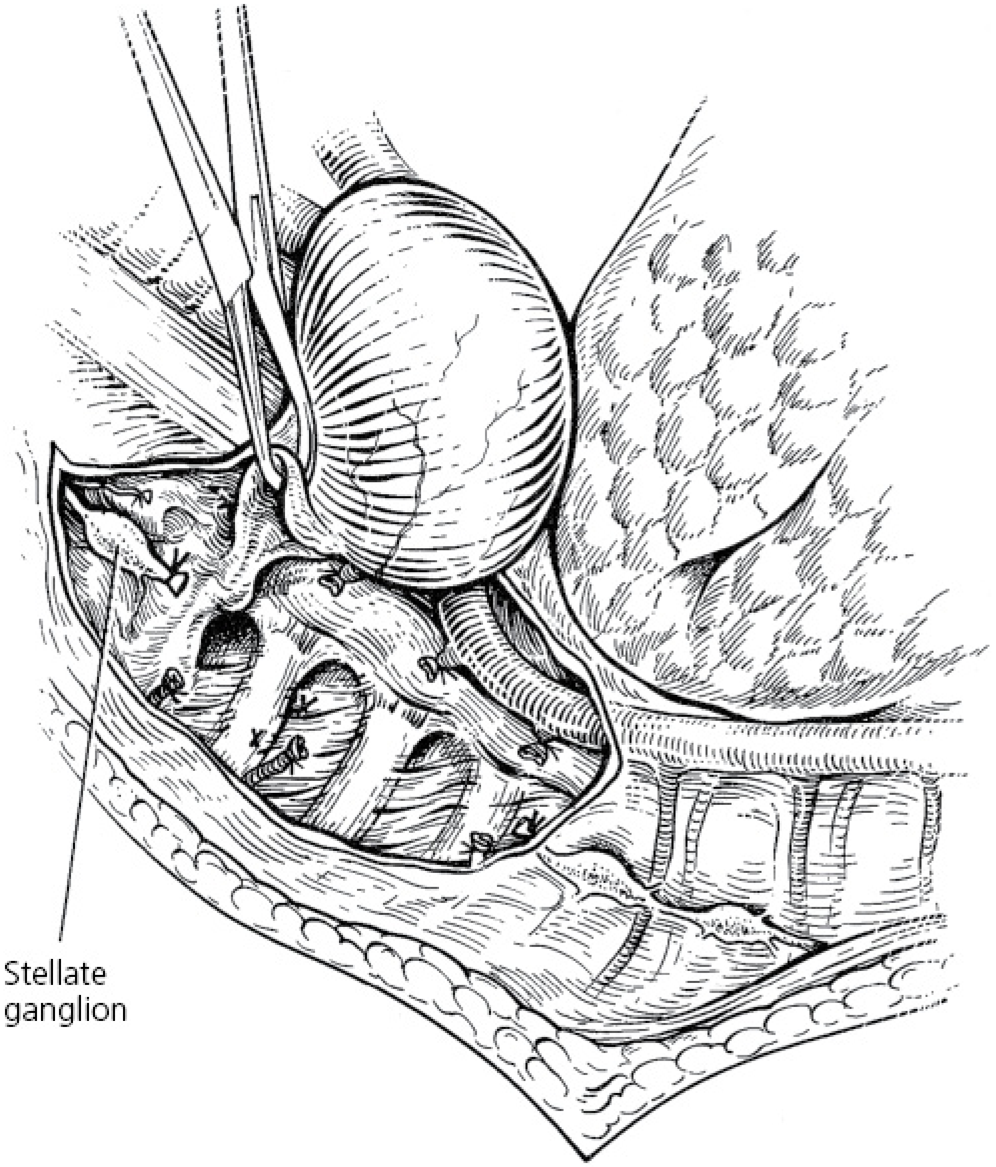
19.11

The most difficult part of this dissection occurs at the sulcus, where the tumor may extend into the neural foramina (**Figure 19.12**). An artery and vein accompany the nerve from each foramen. The use of bipolar cautery or ultrasonic shears in this area avoids the risk of conduction of the current to the spinal cord. The aorta and esophagus, when involved, can generally be dissected from the anterior aspect of the tumor easily as direct involvement of the tumor is rare. As the aorta or azygos vein are dissected forward, each of the intercostal arteries should be controlled and ligated or clipped or cauterized for thoracoscopic resections. The tumor rarely extends through the periosteum of the vertebral bodies. A combination of blunt and cautery dissection is utilized to mobilize the tumor off each of the vertebral bodies.



19.12

Dissection proceeds around the mass, working where progress can most readily be achieved. The sympathetic chain is divided superior and inferior to the tumor. The stellate ganglion should be preserved if it is distinct from the mass to avoid Horner's syndrome (**Figure 19.13**). Ganglioneuromas and neurofibromas do not have an extensive blood supply and are firm and fibrous and easy to keep intact during the dissection. Neuroblastomas are much softer and more vascular, and rupture should be avoided. If preliminary chemotherapy is administered, the neuroblastoma becomes much firmer and also less vascular. A nasogastric tube in the esophagus will facilitate its identification and dissection from a large tumor.



19.13

Once the mass, whether cystic or solid, has been completely resected, the area should be inspected for any ongoing bleeding or evidence of lymphatic leak. These should be controlled and an intercostal chest tube inserted. While oxidized regenerated cellulose (Surgicel®) may be used to help control bleeding in the chest cavity or foramina, it should not be left in the pleural space when the neural foramina have been dissected during the resection as it has been reported to migrate into the spinal canal and produce compression of the cord as the material expands.

Wound closure

It is rarely possible to cover the defect with pleura, particularly following resection of a solid tumor where a significant amount of pleura is removed. A chest tube is placed in the mid-axillary line two intercostal spaces below the incision so it can be brought up through the muscle in a superior trajectory. It can also be brought out at an appropriately positioned port site. It is secured with non-absorbable sutures. For straightforward thoracoscopic procedures requiring minimal dissection, omission of a chest tube can be considered. The ribs are then approximated with pericostal sutures and each of the musculofascial layers is reapproximated with absorbable sutures. Subcuticular polyglycolic acid absorbable sutures are used to reapproximate the skin edges. For the thoracoscopic port sites, absorbable sutures are used to approximate the musculofascial layer and the dermis.

POSTOPERATIVE CARE

Management of the airway

Most children can be extubated following surgery unless the procedure has been particularly long or required significant volume replacement. In infants and children, a humidified oxygen tent will often be better tolerated than a face mask. In very small infants, a short period of postoperative ventilation will allow equilibration and progressive withdrawal of respiratory support. If a child is to remain intubated, an “air leak” must exist around the endotracheal tube; this confirms that the uncuffed tube is not producing undue pressure on the tracheal or subglottic mucosa with a risk of developing a postoperative stricture.

Intercostal drainage

Suction is employed for 24–48 hours and, if drainage is insignificant, the tube can then be removed. An air leak is rare following these resections. The intercostal tube is removed once it is confirmed that there is full expansion of the lung with no air leak. Serous drainage is common, but it does not usually require extended intercostal drainage.

Analgesia and sedation

Young children and adolescents require analgesia for the first several days after surgery. For major resections through a full posterolateral thoracotomy, an epidural catheter or paravertebral and/or intercostal nerve blocks may provide optimum pain relief, particularly in adolescents. Infants require a shorter duration of analgesia. Sedation is also necessary in all infants and children during mechanical ventilation.

FURTHER READING

Fraga JC, Rothenberg SS, Kiely E, Pierro A. Video-assisted thoracic surgery resection for pediatric mediastinal neurogenic tumors. *J Pediatr Surg* 2012; 47(7): 1349–53.

Hammer GB. Single-lung ventilation in infants and children. *Pediatr Anesth* 2004; 14: 98–102.

Hennington MH, Ulicny KS, Detterbeck FC. Vertical muscle-sparing thoracotomy. *Ann Thorac Surg* 1994; 57: 759–61.

Hirose S, Clifton MS, Bratton B, et al. Thoracoscopic resection of foregut duplication cysts. *J Laparoendosc Adv Surg Tech* 2006; 16: 526–9.

Lacreuse I, Valla JS, de Lagausie P, et al. Thoracoscopic resection of neurogenic tumors in children. *J Pediatr Surg* 2007; 42: 1725–8.

Meehan JJ, Sandler AD. Robotic resection of mediastinal masses in children. *J Laparoendosc Adv Surg Tech* 2008; 18: 114–19.

Nordin AB, Fallon SC, Jea A, et al. The use of spinal angiography in the management of posterior mediastinal tumors: Case series and review of the literature. *J Pediatr Surg* 2013; 48: 1871–7.

Partrick DA, Rothenberg SS. Thoracoscopic resection of mediastinal masses in infants and children: An evaluation of technique and results. *J Pediatr Surg* 2001; 36: 1165–7.

Petty JK, Bensard DD, Partrick DA, et al. Resection of neurogenic tumors in children: Is thoracoscopy superior to thoracotomy? *J Am Coll Surg* 2006; 203: 699–703.

Saenz NC, Schnitzer JJ, Eraklis AE, et al. Posterior mediastinal masses. *J Pediatr Surg* 1993; 28: 172–6.

Shamberger RC. Preanesthetic evaluation of children with anterior mediastinal masses. *Semin Pediatr Surg* 1999; 8: 61–8.

Tolg C, Abelin K, Laudenbach V, et al. Open vs thoracoscopic surgical management of bronchogenic cysts. *Surg Endosc* 2003; 19: 77–80.

20

Chest wall deformities

ROBERT E. KELLY JR, MARCELO MARTINEZ-FERRO, AND HORACIO ABRAMSON

INTRODUCTION

Management of chest wall deformities has undergone extensive change in recent years. In some patients, treatment with non-surgical methods is possible both for pectus excavatum and pectus carinatum. Surgical alternatives to established open procedures have become widely accepted. Choosing the correct approach for each patient becomes a central question for the surgeon. Surgical technique is very important for obtaining good results in these patients.

PECTUS EXCAVATUM

PRINCIPLES AND JUSTIFICATION

Pectus excavatum is the most common chest wall deformity in infants, children, and adolescents. Its incidence is estimated at 1 in 400 live births and has a male to female ratio of 4 : 1. The etiology of pectus excavatum is unknown. It has a genetic predisposition with patients having a family history of chest wall deformities in 35–40% of cases. Excavatum patients may have a family history of a carinate or protrusion deformity. The pectus excavatum depression is created by two components: Sternum and costal cartilages, with different morphology in different patients. The pectus excavatum depression may be symmetric or asymmetric. In the asymmetric deformities, the more acute and severe depression is more commonly on the right side. The rarest configuration of pectus excavatum is a combination of ipsilateral depression and a contralateral carinate protrusion. In some series, 90% of cases are noted within the first year of life, but in others the lesion is noted more commonly after the onset of the pubertal growth spurt. The great majority of carinate deformity is noted after the pubertal growth spurt.

Indications

About two-thirds of children with pectus excavatum present with symptoms of shortness of breath during exercise and rapid development of fatigue. Symptomatic improvement after repair is frequently noted. Multiple studies have demonstrated a "restrictive" defect with a decrease in the vital capacity, total lung capacity, and maximum breathing capacity. Despite the symptomatic relief often seen in patients and improvement in their exercise tolerance, the pulmonary function tests are not consistently improved after repair. Several studies have suggested that the symptomatic improvement results from enhanced cardiac function with increased stroke volume achieved by resolution of the anterior compression of the right ventricle following surgical repair. It has also been demonstrated that the chest wall deformity, as well as the physiologic components, can have a significant adverse psychologic impact on patients. Therefore, consideration for surgical repair must include both the physiologic and psychologic aspects of this congenital deformity. Children and their parents must be apprised of the risks and benefits of the surgery prior to proceeding with repair.

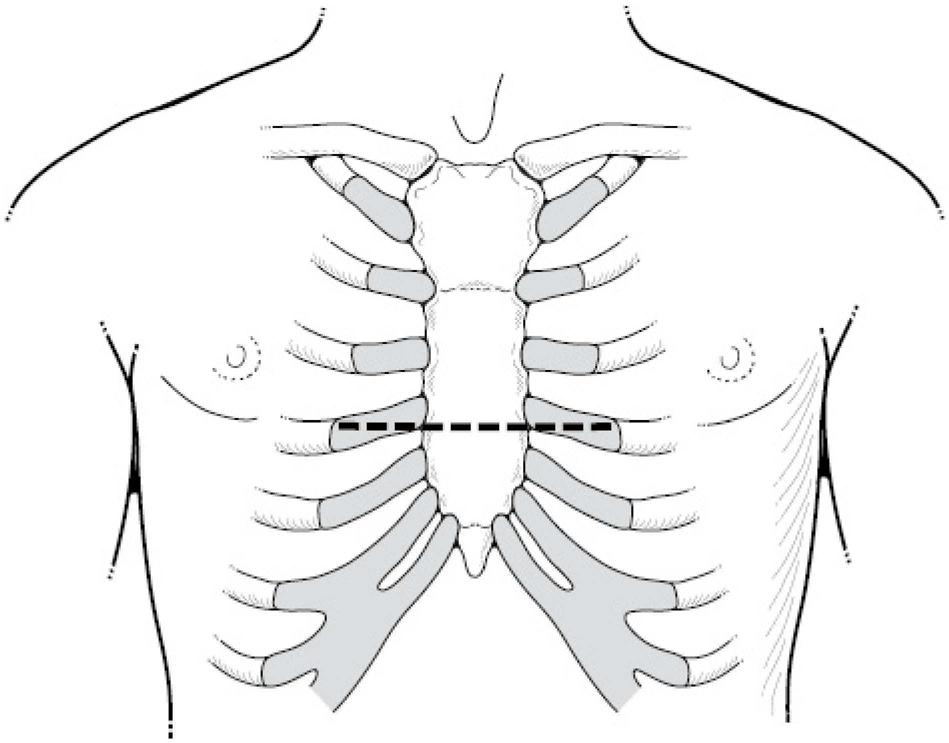
Use of the vacuum bell or suction cup to treat pectus excavatum has been most successful in children younger than 10 years with depressions less than 15 mm, but it has been successful in some older children and adults as well. As worldwide experience accumulates, it will become easier to discriminate which patients will benefit most from this non-invasive treatment. However, even those who fail to obtain correction with vacuum bell therapy have a more pliable chest after treatment, which often improves the overall chest wall contour when surgical treatment is undertaken.

OPERATIONS

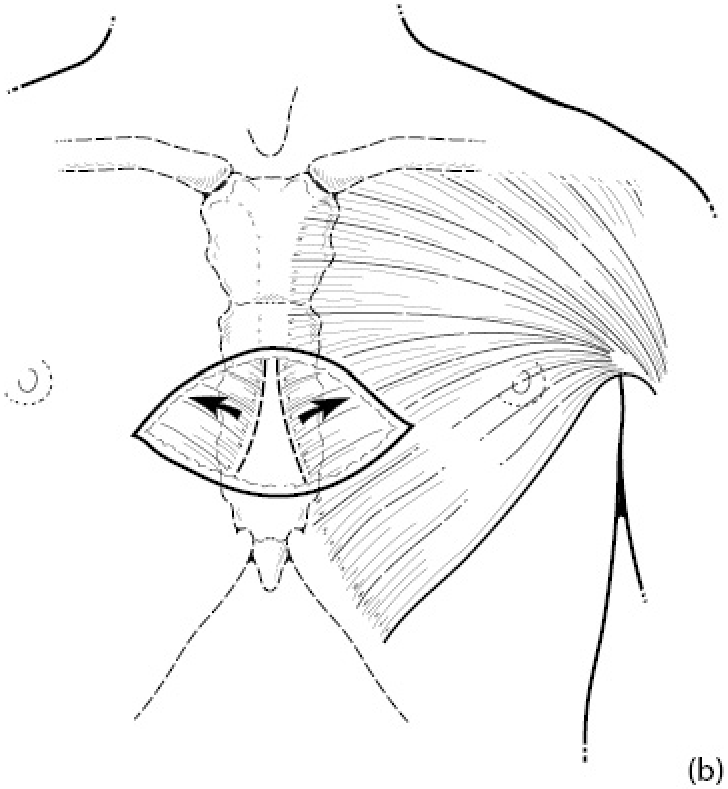
Multiple repairs have been developed for correction of pectus excavatum, but none has been uniformly accepted as the optimal procedure. The Nuss repair presently is most widely utilized. None of the alternatives is optimal in every circumstance. The current standard open repair is frequently attributed to Ravitch. His initial description included resection of the costal cartilage and the perichondrium with anterior fixation of the sternum with Kirschner wires. Welch and Baronofsky subsequently stressed the vital importance of preservation of the perichondrium to achieve optimal regeneration of the cartilage after repair, which generally occurs within 3 months of operation.

Open repair

In the standard open procedure, a transverse incision is made below and well within the nipple lines and, in females, at the inframammary crease (**Figure 20.1a**). The pectoralis major muscle is elevated from the sternum along with portions of the pectoralis minor and serratus anterior bundles (**Figure 20.1b**).



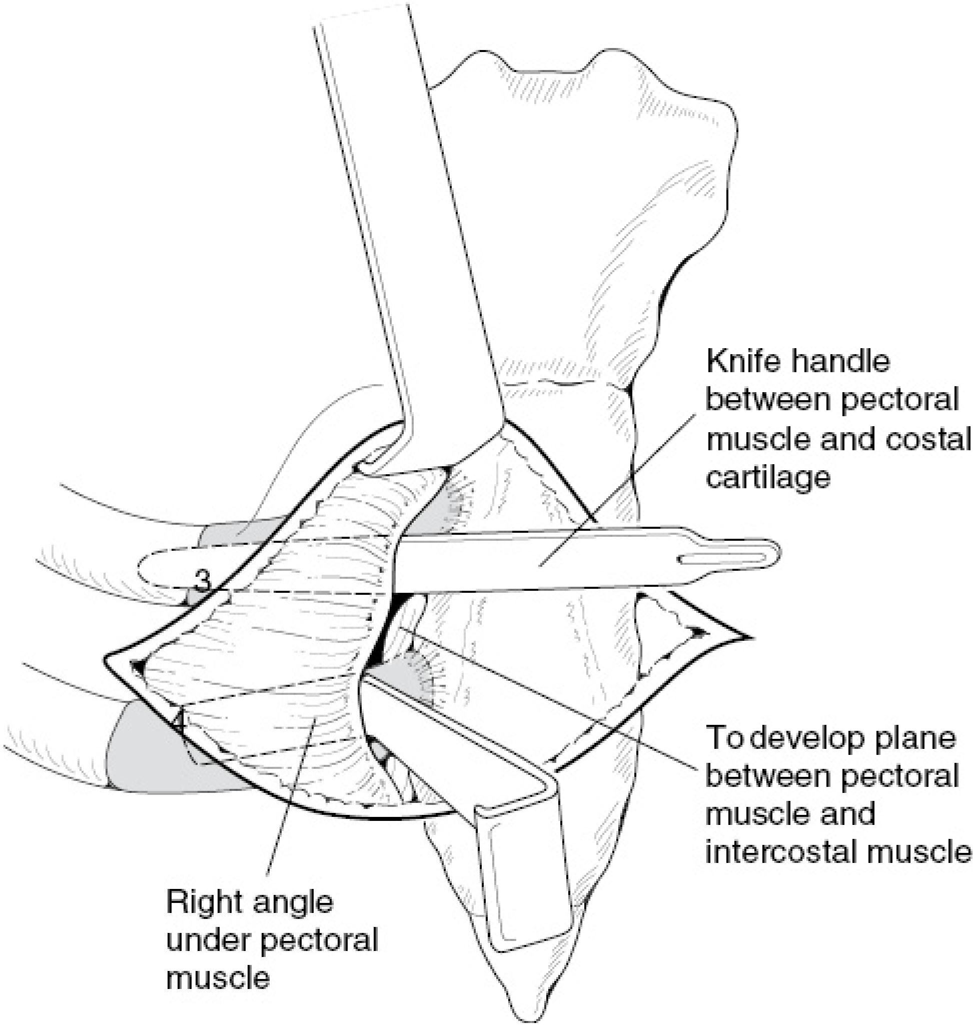
(a)



(b)

20.1a,b

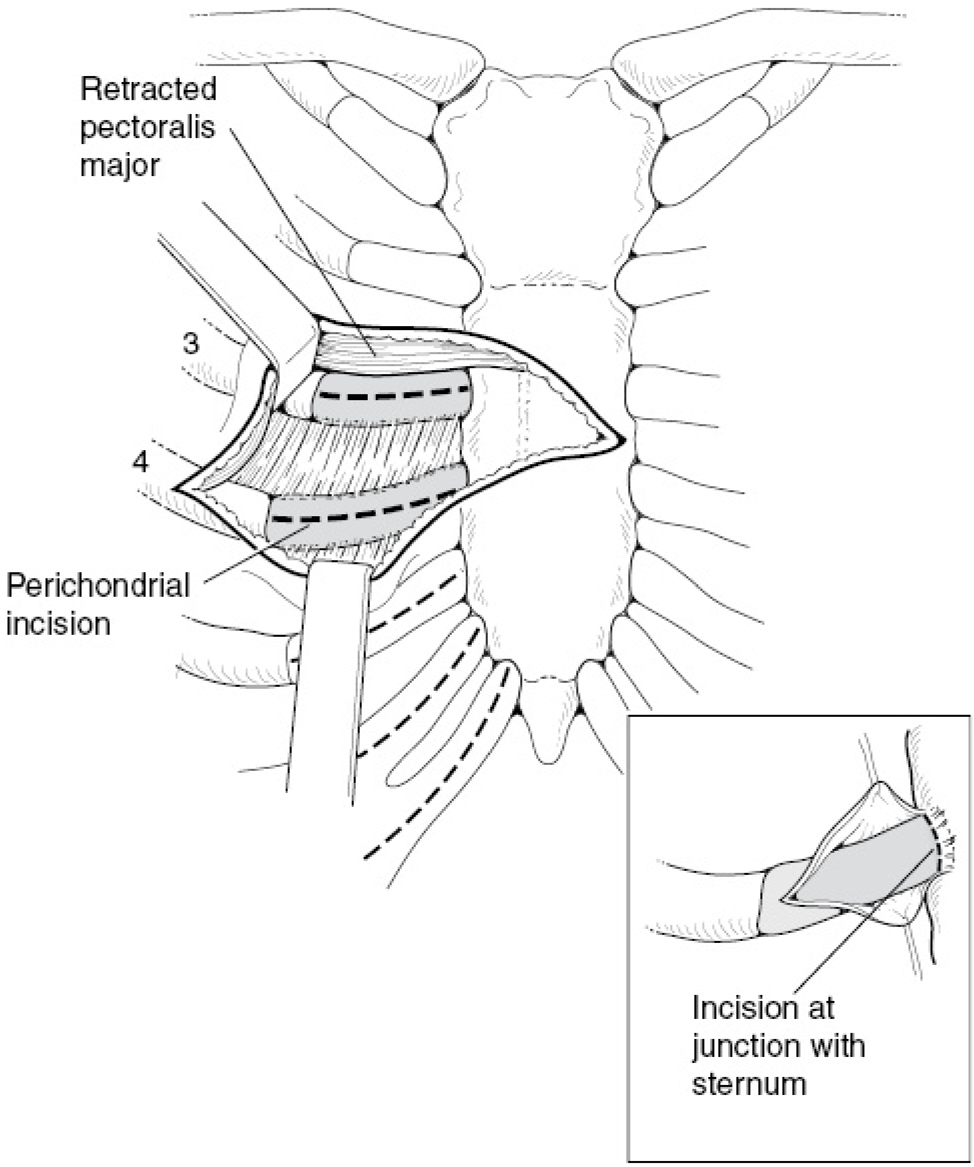
The correct plane of dissection of the pectoral muscle flap is defined by passing an empty knife handle directly anterior to a costal cartilage after the medial aspect of the muscle has been elevated with electrocautery (**Figure 20.2**). The knife handle is then replaced with a right-angle retractor, which is pulled anteriorly. The process is then repeated anterior to an adjoining costal cartilage. Anterior distraction of the muscles during the dissection facilitates identification of the avascular areolar plane and avoids entry into the intercostal muscle bundles. Elevation of the pectoral muscle flaps is extended bilaterally to the costochondral junctions of the third to fifth ribs and a comparable distance for ribs six and seven or to the lateral extent of the deformity.



Knife handle between pectoral muscle and costal cartilage
To develop plane between pectoral muscle and intercostal muscle
Right angle under pectoral muscle
3
4

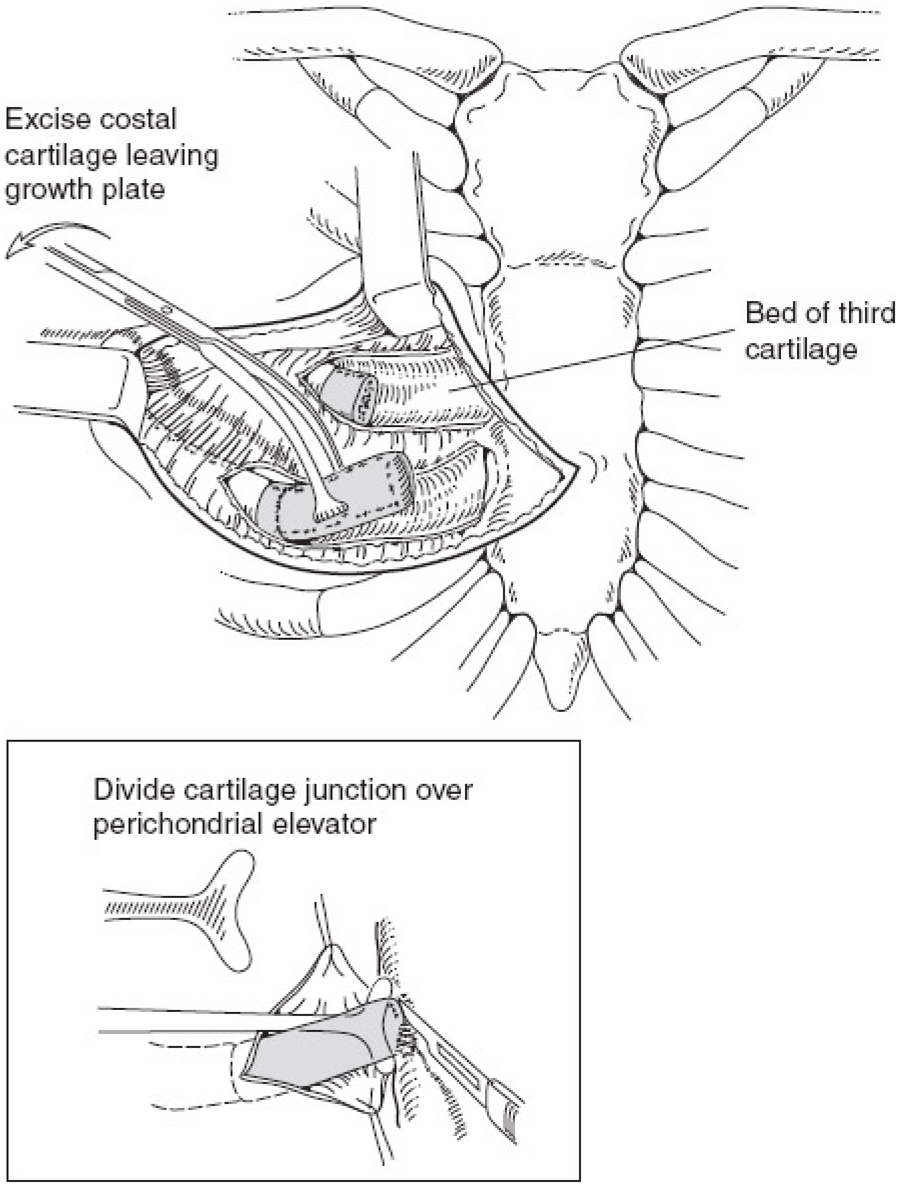
20.2

Subperichondrial resection of the costal cartilages is achieved by incising the perichondrium anteriorly (**Figure 20.3**). The perichondrium is then dissected away from the costal cartilages in the bloodless plane between perichondrium and costal cartilage. Cutting back the perichondrium 90 degrees in each direction at its junction with the sternum (inset) facilitates visualization of the back wall of the costal cartilage.


Retracted pectoralis major
3
4
Perichondrial incision
Incision at junction with sternum

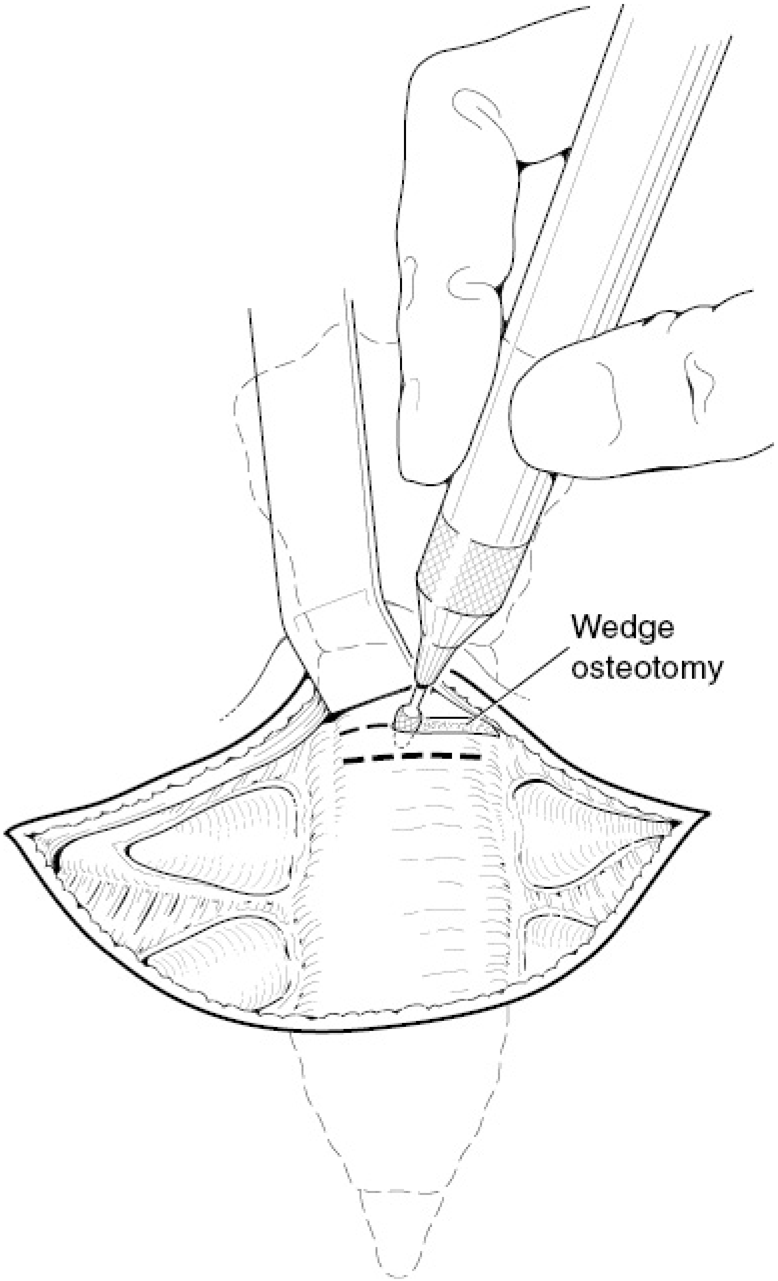
20.3

The cartilages are divided with a knife at their junction with the sternum as a Welch perichondrial elevator is held posterior to the costal cartilage to elevate the cartilage and protect the mediastinum (**Figure 20.4**). The divided cartilage can then be held with an Allis or Lahey clamp and elevated. The costochondral junction is preserved by leaving a segment of costal cartilage on the osseous ribs by incising the cartilage with a scalpel. Fonkalsrud has described resection of segments of the deformed portion of the costal cartilage rather than the entire segment, suggesting that regeneration is improved with this modification. Costal cartilages three to seven are generally resected, but occasionally the second costal cartilages must be removed if posterior displacement of the sternum extends to this level. Segments of the sixth and seventh costal cartilages are resected to the point where they flatten to join the costal arch. Familiarity with the cross-sectional shape of the medial ends of the costal cartilages facilitates their removal. The second and third cartilages are broad and flat, the fourth and fifth are circular, and the sixth and seventh are narrow and deep.


Excise costal cartilage leaving growth plate
Bed of third cartilage
Divide cartilage junction over perichondrial elevator

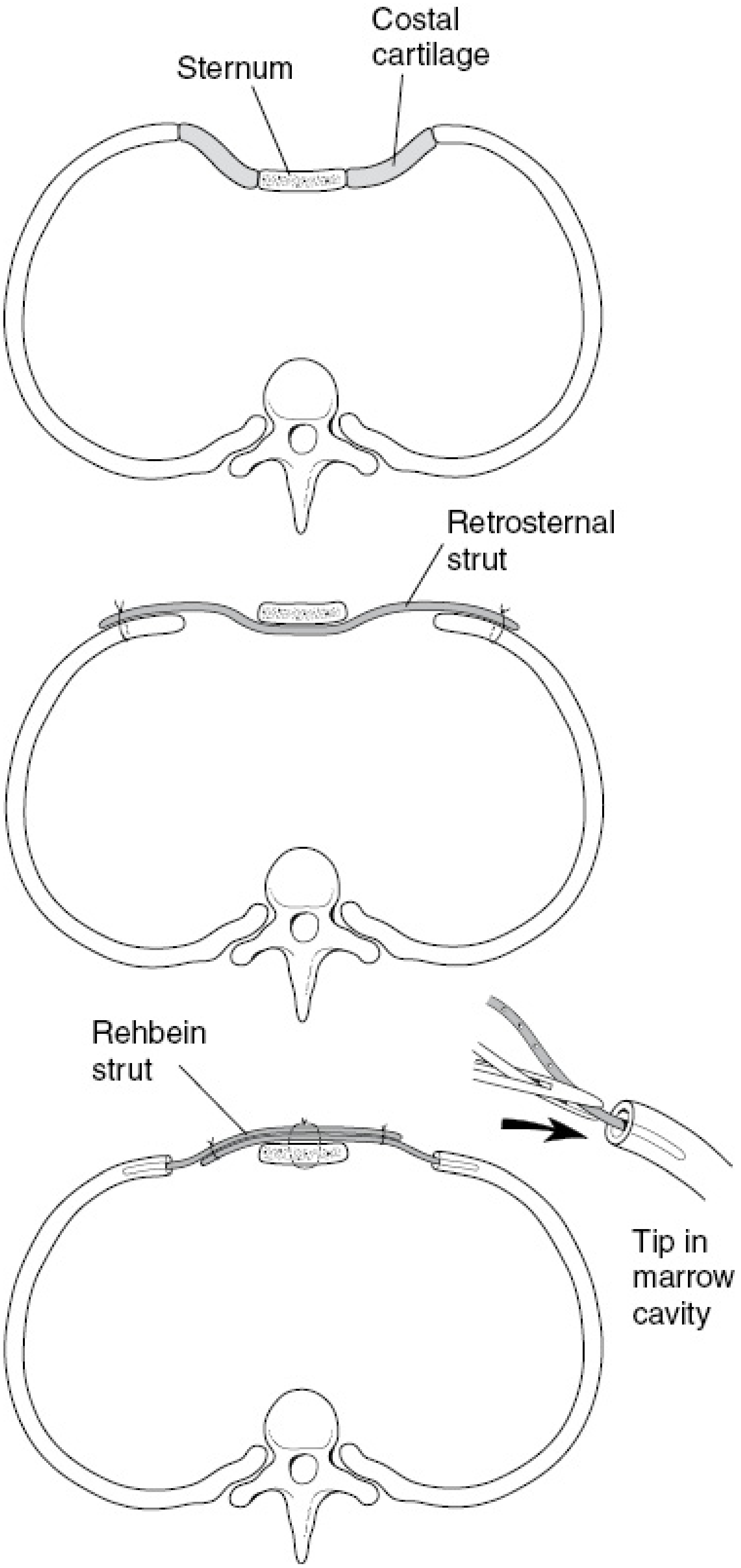
20.4

The sternal osteotomy is created at the level of the posterior angulation of the sternum, generally at the level of the insertion of the third cartilage, but occasionally the second. Two transverse sternal osteotomies are created through the anterior cortex with a Hall air drill 3–5 mm apart, and the wedge of bone is partially mobilized (**Figure 20.5**). The base of the sternum and the rectus muscle are elevated with two towel clips, and the posterior plate of the sternum at the osteotomy is fractured. The xiphoid can be divided from the sternum if its anterior angulation produces an unsightly bump below the sternum when it is elevated in its corrected position. The insertion of the rectus muscle into the sternum can generally be preserved by dividing the xiphoid with electrocautery by a lateral approach. Preservation of the attachment of the perichondrial sheaths and xiphoid to the sternum avoids an unsightly depression that can occur below it. When a strut is not used, the osteotomy is closed with several heavy silk sutures as the sternum is elevated by the assistant.



20.5

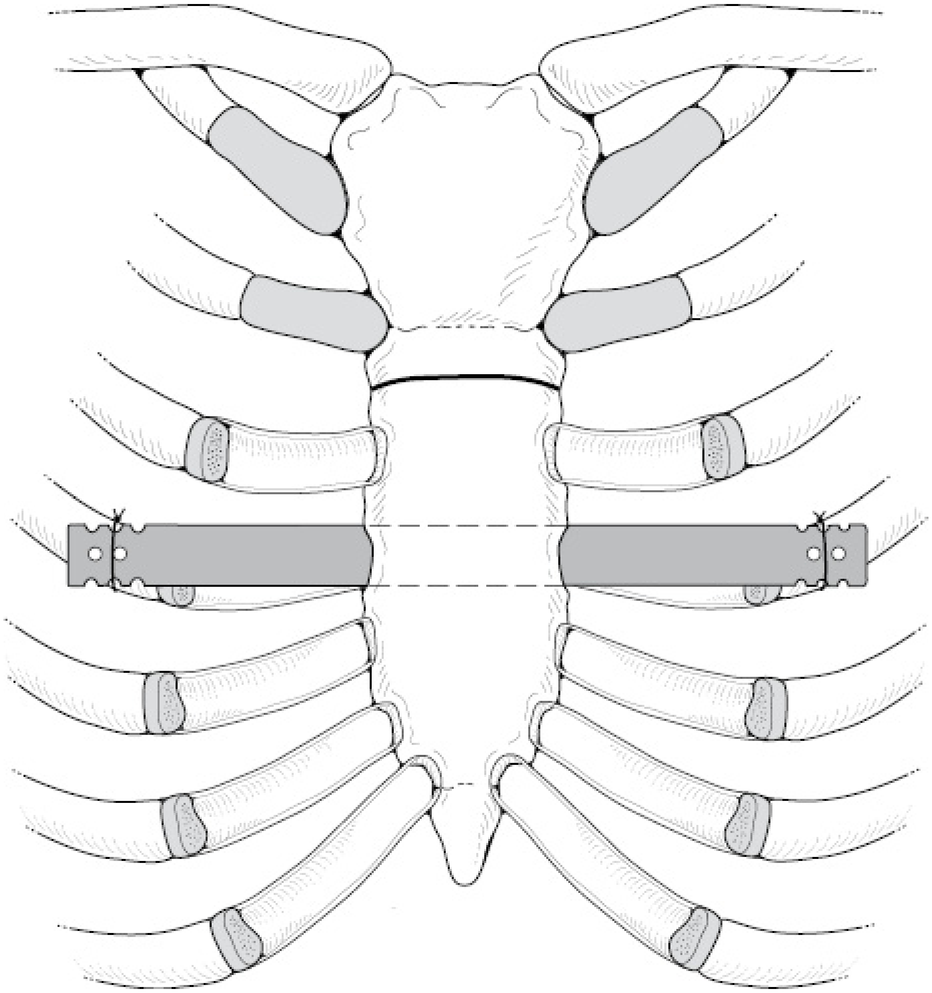
Figure 20.6 demonstrates the use of both retrosternal struts and Rehbein struts. Rehbein struts are inserted into the marrow cavity (insert) of the third or fourth ribs, and the struts are then joined medially with stainless steel wire to create an arch anterior to the sternum. The sternum is sewn to the arch to secure it in its corrected position. The retrosternal strut is placed behind the sternum and is secured to the rib ends laterally to prevent migration.



Costal cartilage
Sternum
Retrosternal strut
Rehbein strut
Tip in marrow cavity

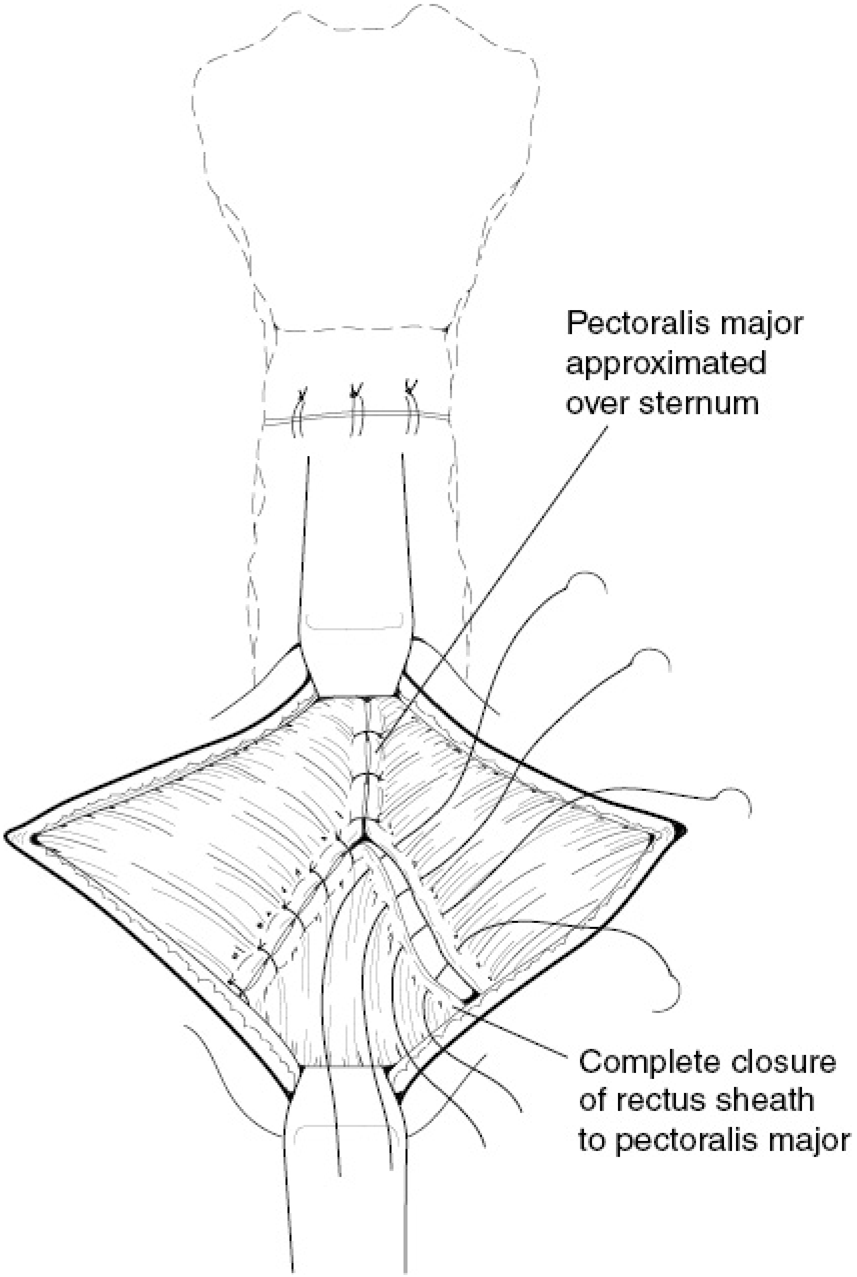
20.6

Figure 20.7 is an anterior depiction of the retrosternal strut. The perichondrial sheath to either the third or fourth rib is divided from its junction with the sternum, and the retrosternal space is bluntly dissected to allow passage of the strut behind the sternum. It is secured with two pericostal sutures at each end to prevent lateral migration. The wound is then flooded with warm saline and cefazolin solution to remove clots and inspect for a pleural entry. A single-limb medium Hemovac drain is brought through the inferior skin flap and placed in a parasternal position.



20.7

The pectoral muscle flaps are secured to the midline of the sternum, advancing the flaps inferiorly to obtain coverage of the entire sternum. The rectus muscle fascia is then joined to the pectoral muscle flaps, closing the mediastinum (**Figure 20.8**).


Pectoralis major approximated over sternum
Complete closure of rectus sheath to pectoralis major

20.8

POSTOPERATIVE CARE

Perioperative antibiotics are utilized by most surgeons. Pain control is managed by either an epidural catheter or IV narcotics in the immediate perioperative period. Most surgeons utilize closed drainage early after repair to minimize the risk of hematoma or seroma development under the flaps. Children are encouraged to ambulate early after repair. Contact sports are generally avoided during the first 6 months of repair with either suture or strut elevation of the sternum.

OUTCOME

Generally successful repair of pectus excavatum is reported. It is critical that children are followed until they reach their full stature for it is during the period of rapid growth during puberty that recurrences may occur. No randomized studies between sternal fixation with struts and suture fixation have been performed. Comparable outcomes are reported in large series using both techniques. One long-term series from a single institution, however, reports improved outcome in those patients with retrosternal strut fixation.

Minimally invasive repair: Nuss technique

INDICATIONS

A complete history and physical examination is performed on all patients and includes documenting photographs. Patients who have a mild to moderate pectus excavatum are treated with an exercise and posture program in an attempt to halt progression of the deformity. The exercises are designed to improve posture, strengthen the chest and back muscles, and improve exercise tolerance. These patients are followed at 6- to 12-month intervals.

Patients who have a severe deformity or who have documented progression are also treated with the exercise and posture program, but, in addition, they undergo objective studies to see whether their condition is severe enough to warrant surgical correction. These studies include a thoracic computed tomography (CT) scan, pulmonary function tests, and a cardiac evaluation that includes an electrocardiogram (ECG) and an echocardiogram. The use of magnetic resonance imaging (MRI) can easily be substituted for a CT scan.

Determination of a severe pectus excavatum and the need for repair requires two or more of the following criteria:

1. A Haller CT index greater than 3.25, or correction index greater than 10%
2. Pulmonary function studies that indicate restrictive airway disease
3. A cardiology evaluation demonstrating the compression is causing murmurs, mitral valve prolapse, cardiac displacement, or conduction abnormalities on the echocardiogram or ECG tracings
4. Documentation of progression of the deformity with associated physical symptoms other than isolated concerns of body image.

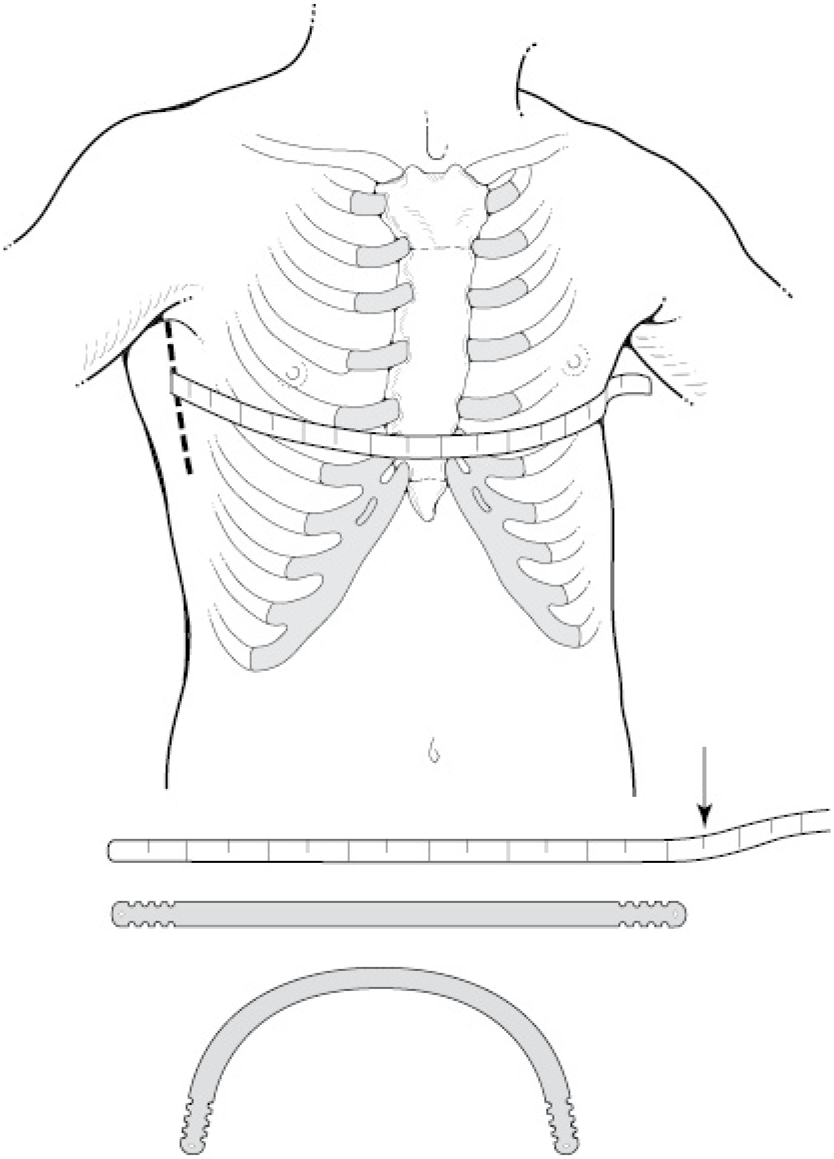
When utilizing these criteria, less than 50% of patients are found to have a deformity severe enough to warrant surgery.

Our experience has shown that the optimal age for repair is 10–14 years, as the patients' chests are still soft and malleable and they show rapid recovery and return to normal activities, and have excellent structural results. After puberty, the flexibility of the chest wall is decreased, which often necessitates the insertion of two bars. It also takes the patients longer to recover when they are older. However, we have performed the procedure in patients up to age 48 years with equally good long-term results.

TECHNIQUE

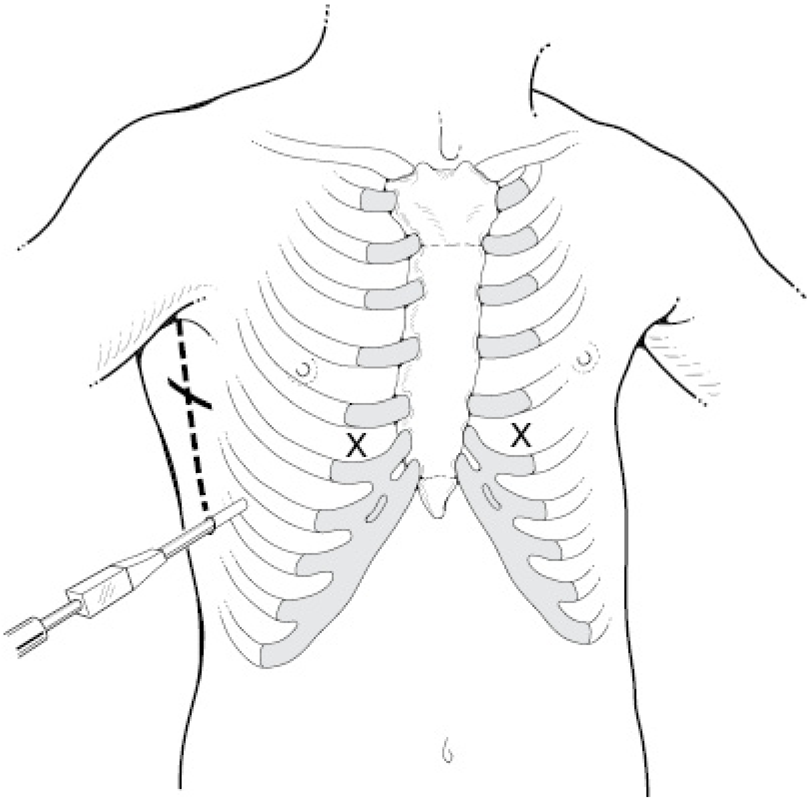
A first-generation cephalosporin is used for antibiotic coverage and continued for 24 hours. The arms are abducted 75 degrees at the shoulders, the elbows slightly flexed, and rolled up foam is placed beneath the wrists and hands to keep the upper extremities relaxed and without tension on the brachial plexus.

The length of the pectus bar is determined by measuring the distance from the right mid-axillary line to the left mid-axillary line and subtracting 2.5 cm, because the bar takes a shorter course than the tape measure. The measurement is done over the area of the deepest depression that is still part of the sternum. The bar is bent to the desired convex configuration, bearing in mind that the center of the bar should be flat for 2–4 cm to provide greater stability (**Figure 20.9**). Use of a second or even third bar should be considered in older patients, in those with longer or deeper depressions, and in those with a stiff chest. At present we utilize two bars in more than half our patients. Two bars distribute the load over a greater area, and by lifting up a greater area can improve the overall thoracic contour.



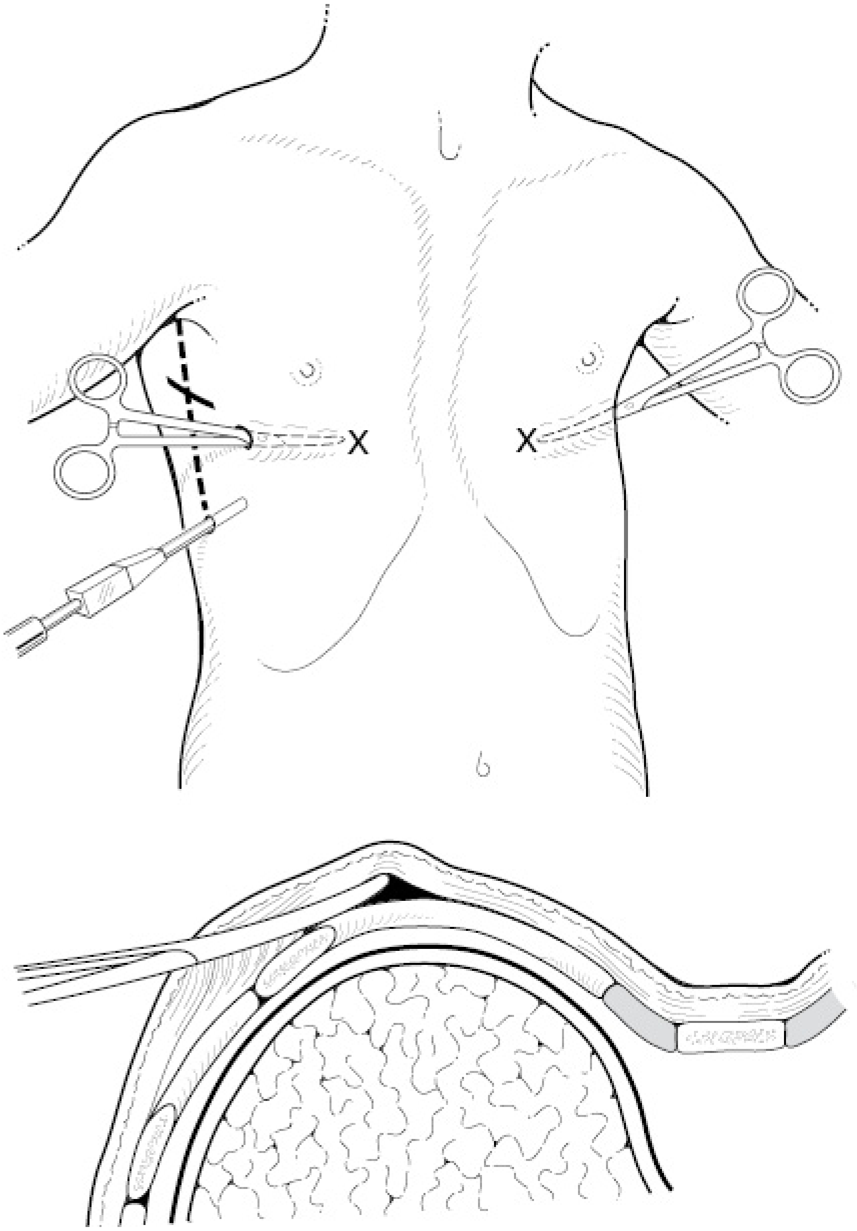
20.9

A 30-degree, 5 mm thoracoscope is inserted into the right chest two intercostal spaces below the site of planned bar placement to confirm that the internal anatomy corresponds with the external markings and to look for unexpected pathology. If all is well, then transverse lateral thoracic incisions are made between the mid- and anterior axillary lines and subcutaneous tunnels are created to the greatest apex of the pectus deformity which is marked with an ”X” (**Figure 20.10**). These Xs represent the entrance and exit sites of the bar from the chest. They are in the intercostal space that is in the same horizontal plane as the deepest depression marked with a circle and care should be taken that they are placed medial to the greatest apex of the chest.



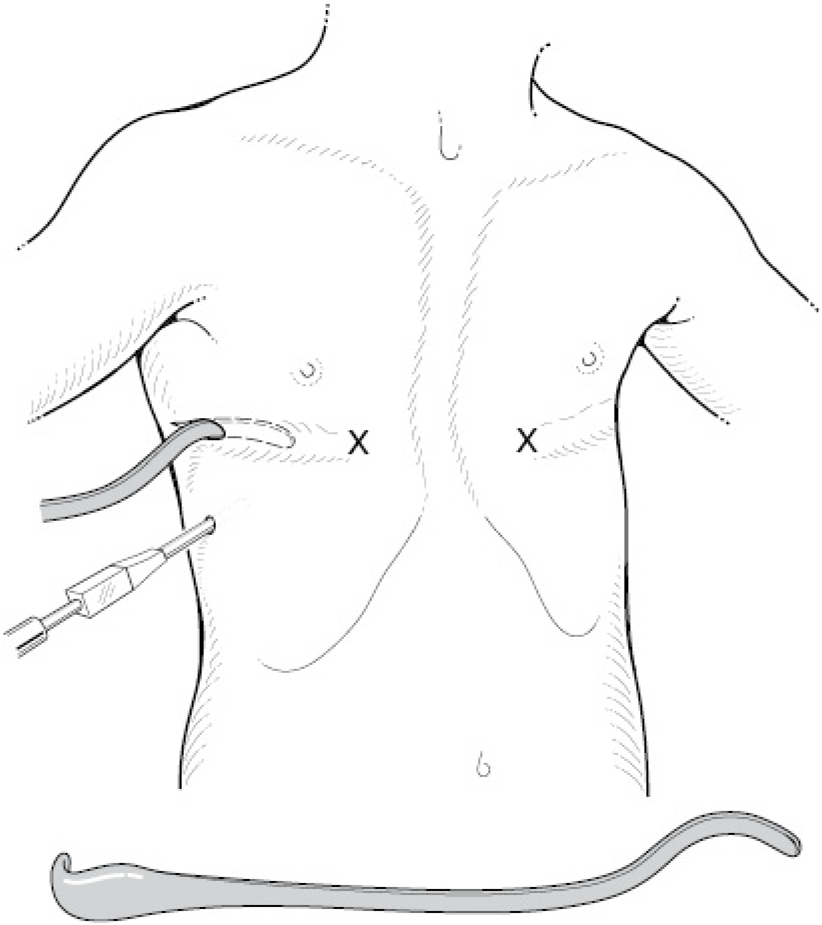
20.10

Skin tunnels are created above the muscle starting from each of the lateral thoracic incisions to the top of the pectus ridge on each side. The tunnel can also be created between the pectoralis major and minor muscles to minimize skin retraction (**Figure 20.11**). The tunnels should be created so that the entry and exit sites of the bar from inside the chest are medial to the top of the pectus ridge on each side. With the thoracoscope in place, a tonsil clamp is inserted into the subcutaneous tunnel on the right and a blunt thoracostomy is created at the site marked “X,” taking care not to injure the intercostal vessels, lung, or pericardium.



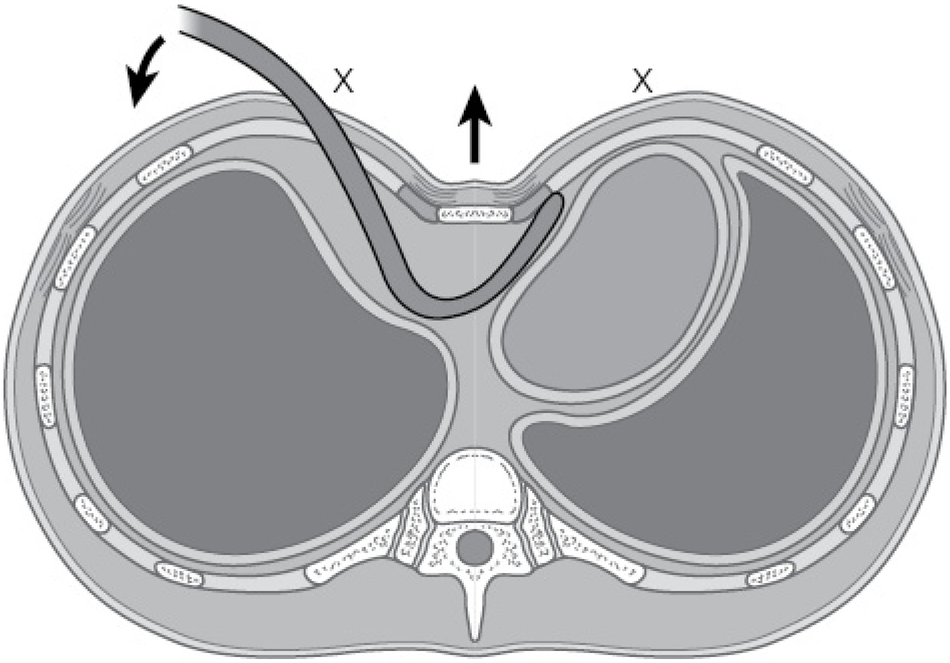
20.11

Under continuous thoracoscopic visualization, a Biomet™ (Biomet Microfixation, Jacksonville, FL, USA) introducer is inserted into the chest through the right tunnel and thoracostomy site at the top of the pectus ridge (**Figure 20.12**). With great care and thoracoscopic guidance, the pleura and pericardium are dissected off the undersurface of the sternum creating a substernal tunnel. The introducer tip is kept in view at all times and slowly advanced across the mediastinum and brought out through the corresponding intercostal space on the left and advanced out of the incision on the contralateral side. Again, this exit site is medial to the top of the pectus ridge.



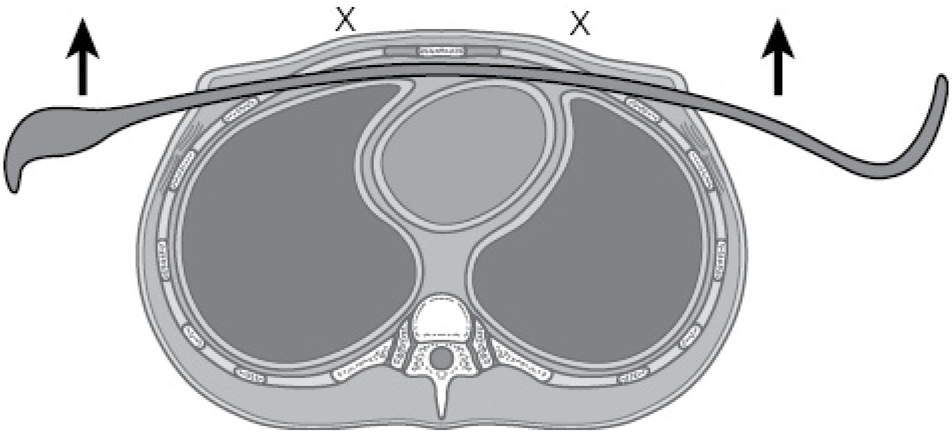
20.12

A 30-degree scope facilitates visualization during the substernal dissection and care is taken to keep the point of the dissector adjacent to the undersurface of the sternum at all times to push the heart out of the way of the dissection plane (**Figure 20.13**). There should be a clear plane between the pericardium and the sternum. During the dissection, the ECG monitor should be turned to maximum volume to listen for ectopy or arrhythmias. A subxiphoid incision and manual elevation of the sternum can be used as an adjunct for those patients with very severe deep depressions where adequate visualization cannot be achieved. We utilize sternal elevation with the Rul-Tract in many cases. In severe deep depressions, it is usually necessary to create two tunnels. By creating the first tunnel more superiorly and leaving the introducer in place, the sternum remains elevated, which makes tunneling under the deepest point of the depression much easier, as well as safer.



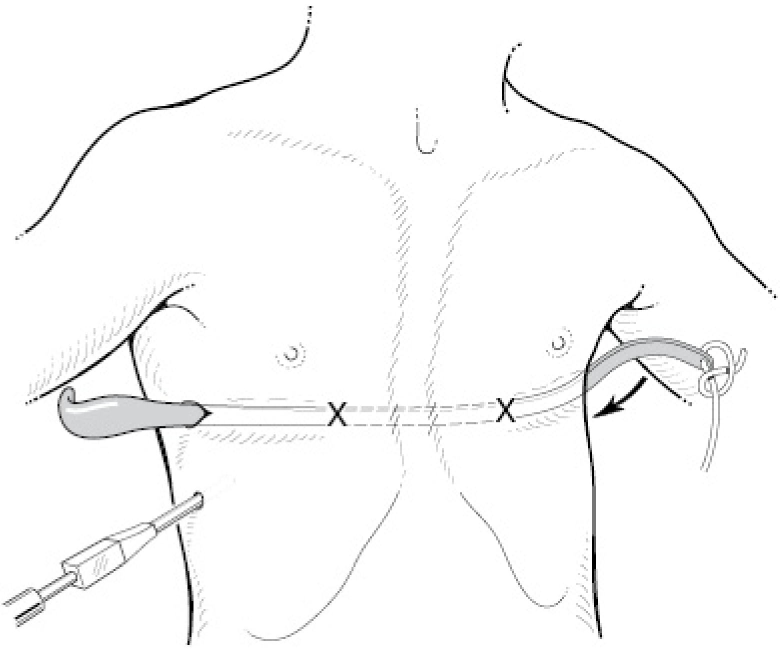
20.13

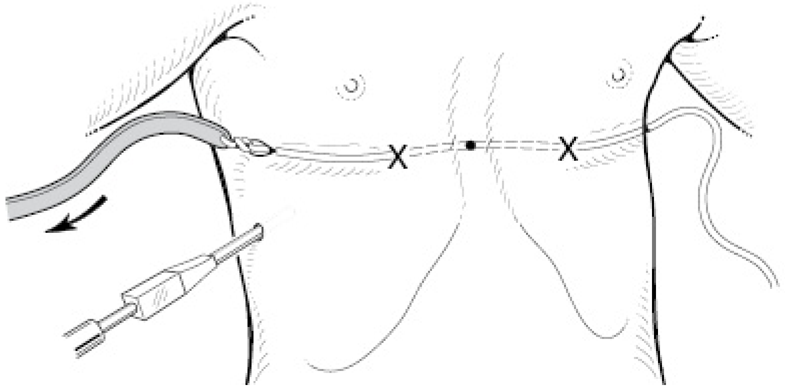
The introducer is pushed out of the thorax through the previously marked intercostal space “X” on the left and advanced out through the corresponding tunnel and incision. When the introducer is fully in place, the sternum is elevated by lifting the introducer on each side, thus correcting the pectus excavatum (**Figure 20.14**). The sternum is lifted out of its depressed position with the introducer numerous times, as well as pressing down on the lower chest wall while lifting the introducer.



20.14

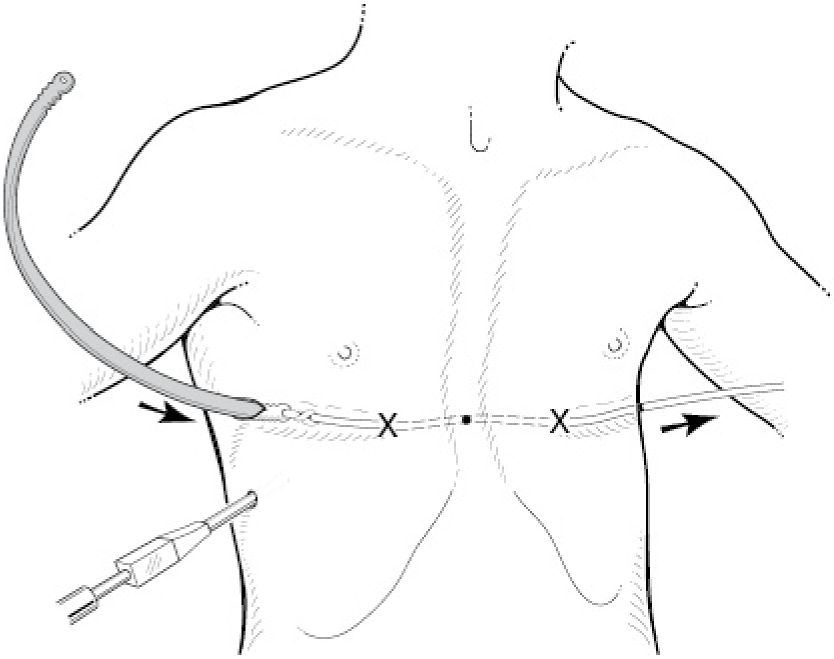
Once the sternal depression has been corrected, an umbilical tape is attached to the introducer and the introducer is slowly withdrawn from the chest cavity with the umbilical tape attached (**Figure 20.15**).





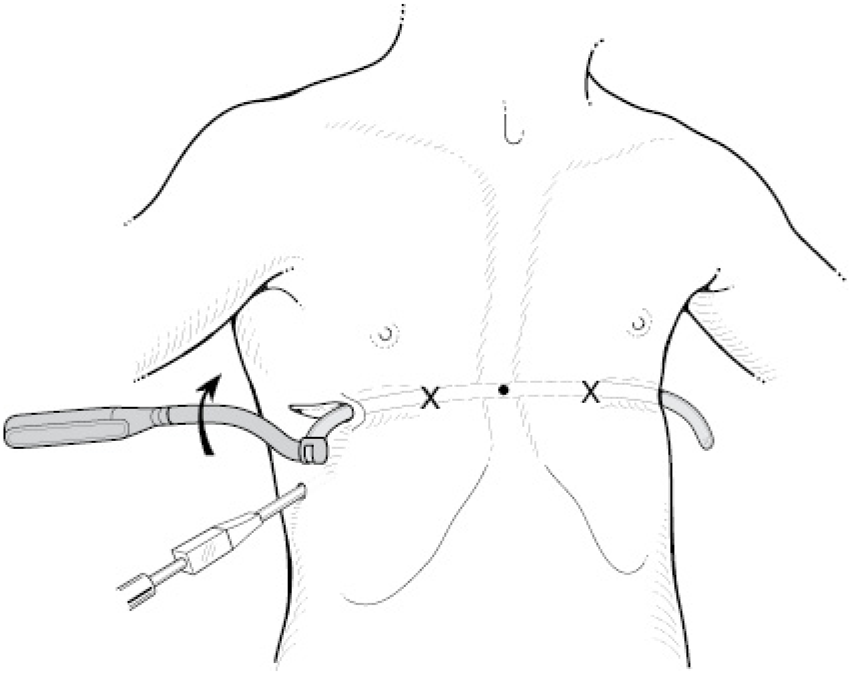
20.15

The pectus bar that was previously bent into a convex shape is then attached to the umbilical tape, slowly guided through the right subcutaneous tunnel under thoracoscopic visualization, and guided through the substernal tunnel with its convexity facing posteriorly until it emerges on the contralateral side (**Figure 20.16**).



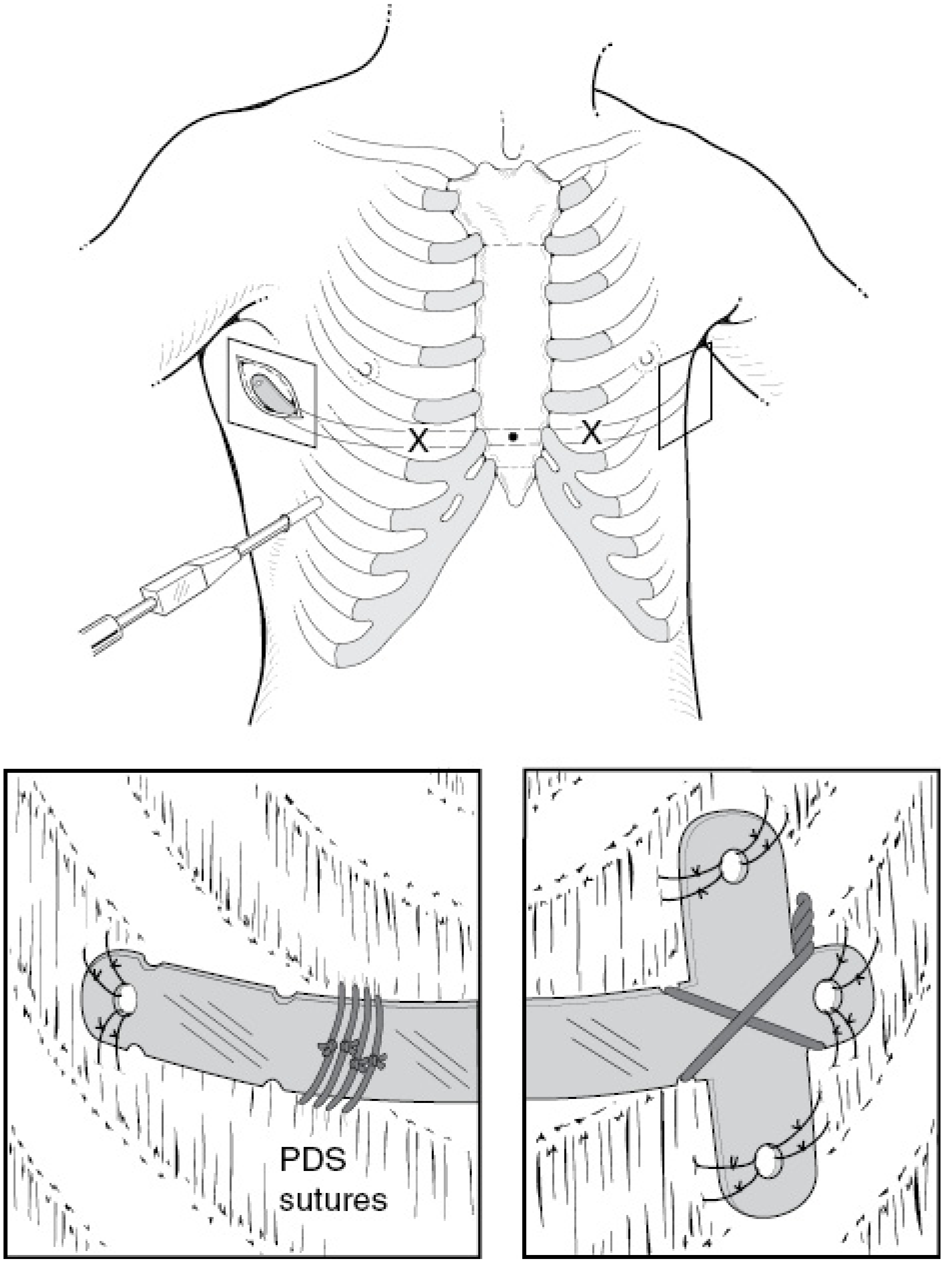
20.16

The pectus bar is positioned inside the chest with its convexity facing posteriorly with an equal amount of bar protruding on each side. Using the specially designed Biomet bar flippers, the bar is rotated 180 degrees, achieving an immediate correction to the pectus deformity (**Figure 20.17**). The sides of the bar should be resting comfortably against the musculature and should not be too tight or too loose. If the bar does not fit snugly on each side because of pressure on the middle, the bar can be reflipped and molded as necessary while still in place in the chest.



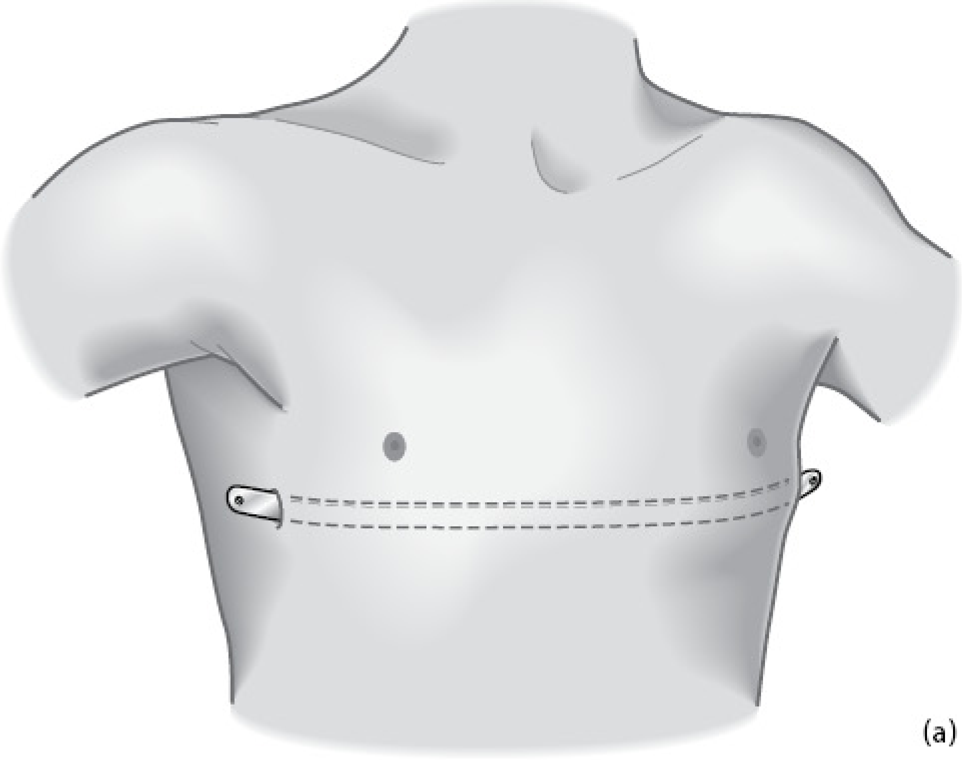
20.17

Bar stabilization is essential for a successful outcome. The bar is stabilized first by attaching a stabilizer to the left end of the bar and wiring the bar and stabilizer together with no. 3 surgical steel wire, or alternatively FiberWire™ (Arthrex, Naples, FL, USA) can be used. If two bars are placed, a stabilizer is placed on each bar on alternating sides. An additional stabilizing technique utilizes a laparoscopic "autosuture" needle to place multiple 0 polidioxanone sutures (PDS) or Vicryl™ sutures around the bar and underlying ribs with thoracoscopic guidance. Ideally, these sutures should be placed around separate ribs for each bar. Since thoracoscopy is usually performed on the right, we place these pericostal sutures on the right and the metal stabilizer on the left (**Figure 20.18**). The stabilizer and bar are also secured by placing numerous interrupted absorbable sutures through the holes in the bar and adjacent fascia.

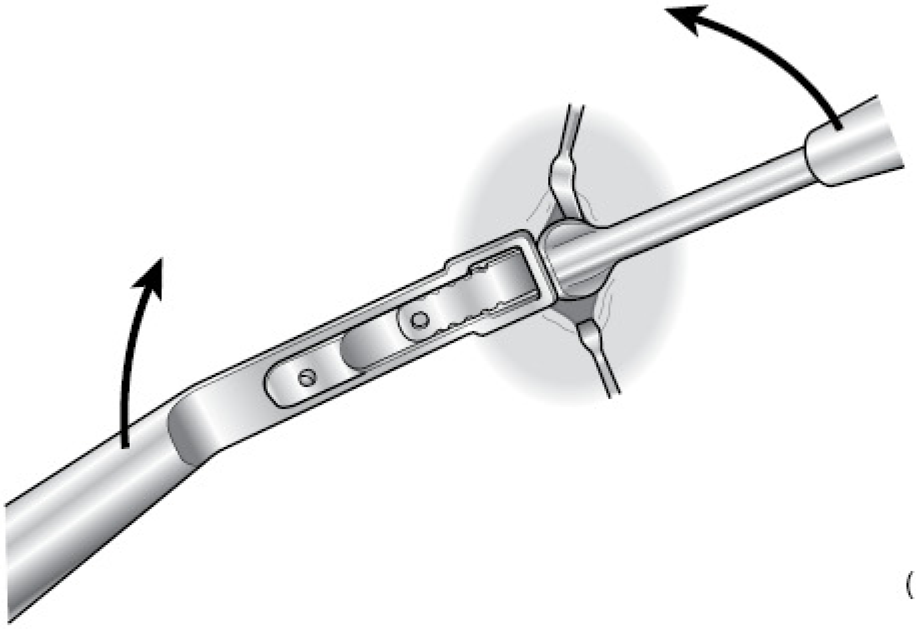


20.18

Bars are removed electively between 2 and 4 years after placement. Bar removal is performed under general anesthesia, using positive end-expiratory pressure (PEEP) to minimize the risk of a pneumothorax. We recommend opening all incisions and mobilizing both ends of the bar (**Figure 20.19a**). It is not uncommon to encounter significant calcifications that require a rongeur and/or chisel to remove. Once the stabilizer is removed, the bars are unbent using Biomet bar flippers or alternatively Biomet Multi-Benders (**Figure 20.19b**). The bars are then removed from the either side gently under positive pressure ventilation using an orthopedic bone hook (**Figure 20.19c**). We routinely perform bar removals as an outpatient procedure and the use of postoperative narcotics is minimal.



(a)



(b)

20.19a–c

POSTOPERATIVE CARE

Perioperative antibiotics are continued for 24 hours. We use patient-controlled analgesia pumps for all patients and transition to oral pain medications on postoperative day 2–3. The patients are discharged on a combination of narcotic, anti-inflammatory, and muscle relaxant medications usually on postoperative day 3. The use of a bilateral ON-Q® Pain Buster® (I-Flow Corporation, Lake Forest, CA, USA) has been a beneficial adjunct to the perioperative pain management. These are placed at the time of operation and usually removed on postoperative day 2.

For the first 6 weeks, patients are prohibited from playing sports, but they are encouraged to do deep-breathing exercises and to ambulate. At 6 weeks post-repair, they are encouraged to resume normal activities, and at 3 months they may resume competitive sports. Heavy contact sports, such as American football, boxing, and ice hockey, are prohibited until the bar has been removed, but other aerobic sports are encouraged.

OUTCOME

In 1997 we published our initial 10-year experience with 42 patients utilizing the minimally invasive technique for pectus repair. In 2010, we published our 21-year experience performing the Nuss repair on 1215 patients. Of those patients, 92 were redo operations, and only 11 of those patients had their primary repair performed at our institution, yielding a less than 1% rate of recurrence overall.

There have been no deaths at our institution and minimal postoperative morbidity. The most common early complication is a pneumothorax, which usually resolves spontaneously and rarely requires a chest tube. The infection rate is less than 2.5%. Bar displacement has been decreased to 1% with the advent of wiring the stabilizers to the bar on the left side, using PDS sutures around the bar and underlying rib on the right side, and the liberal use of two bars on stiff and older patients. These results have continued over the last 10 years at Children's Hospital of The King's Daughters in Norfolk, VA, USA.

PECTUS CARINATUM

PRINCIPLES AND JUSTIFICATION

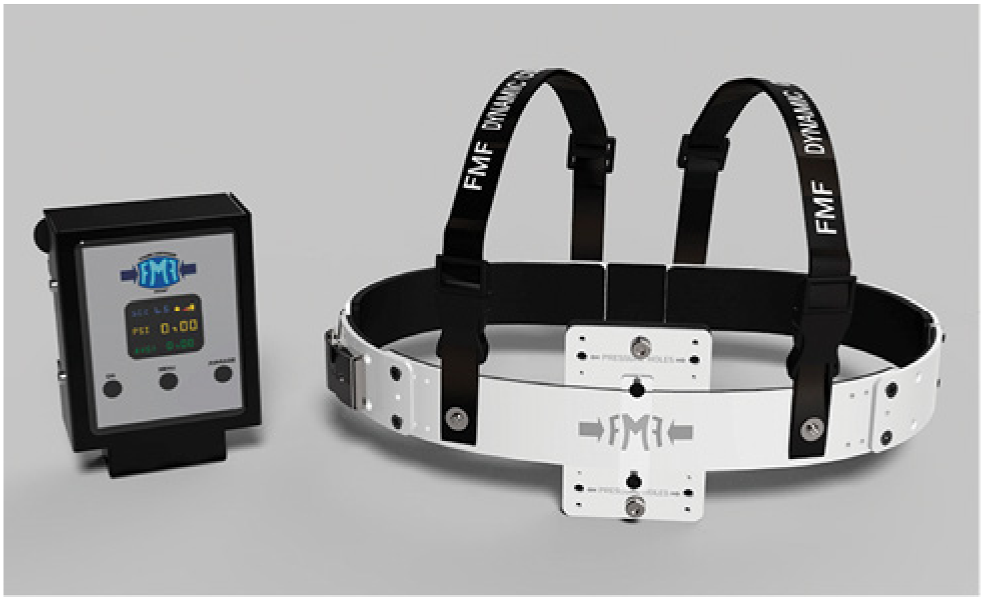
The carinate deformity is much less common than the depression deformity, with approximately a 1:5 ratio of occurrence. It is more common in boys than in girls (4:1), as is pectus excavatum. The deformity frequently appears after the eleventh birthday. In those children in whom a protrusion is noted at birth or in childhood, it often worsens during the period of rapid pubertal growth. The etiology of this deformity is no better understood than is that of pectus excavatum. That a family history is identified in 26% of patients suggests a genetic predisposition. Scoliosis occurs in conjunction with pectus carinatum in 15% of children, which implies a diffuse abnormality in connective tissue development. Consideration for repair of this deformity is based entirely upon the severity of the anterior protrusion. Cardiopulmonary abnormalities are rarely identified.

Correction of the deformity by bracing has been reported and is now the first approach for correction. Compliance with the compression regimen may be an issue. We have utilized several braces over time, and have had by far the best success with the FMF® brace described here over the last 10 years.

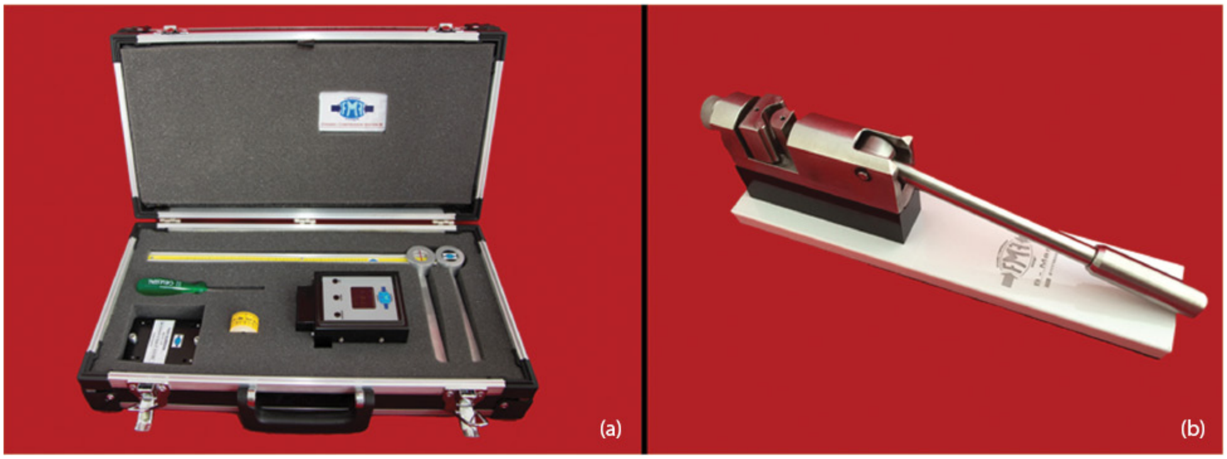
BRACING

The FMF Dynamic Compressor System (DCS) (**Figure 20.20**) comprises the following elements:

- A custom-fitted, expandable, low-profile, cushioned, ultra-light aluminum brace that is adjustable to any thoracic shape or size and has a locking system on one side to keep it in place
- An adaptable compression plate to be placed against the chest wall protrusion
- A tool briefcase (**Figure 20.21**) that contains the following items:



20.20 FMF Dynamic Compressor System. Pressure measuring device (PMD) (left) and custom-made aluminum brace (right).



20.21a,b (a) Briefcase with the measurement elements (PMD, caliper/ruler, metric tape, and screwdriver); (b) portable plate bender.

- A pressure measuring device (PMD) that can be docked to the brace's compression plate and measures the pressure exerted to the patient in pounds per square inches (PSI)
- A chest measuring caliper/ruler
- A metric tape
- A portable plate bender to curve the aluminum segments according to the patient's initial chest size and shape and throughout the continuous reshaping process
- Magnetic screwdrivers and screws

The FMF DCS corrects pectus carinatum gradually by using the continuous anterior–posterior compression to widen and remodel the entire chest and cartilages, which accommodate, grow, and finally ossify in the correct position.

Two measurements were defined by the authors and must be collected:

- *Pressure of correction (PC)* is an indirect parameter of the chest wall's flexibility. It is defined as the pressure applied to the patient, in the most protruding area of the chest, needed to accomplish a proper shape of the thorax.
- *Pressure of treatment (PT)* is the pressure required to treat the patient. It should be measured before and after adjusting the FMF DCS.

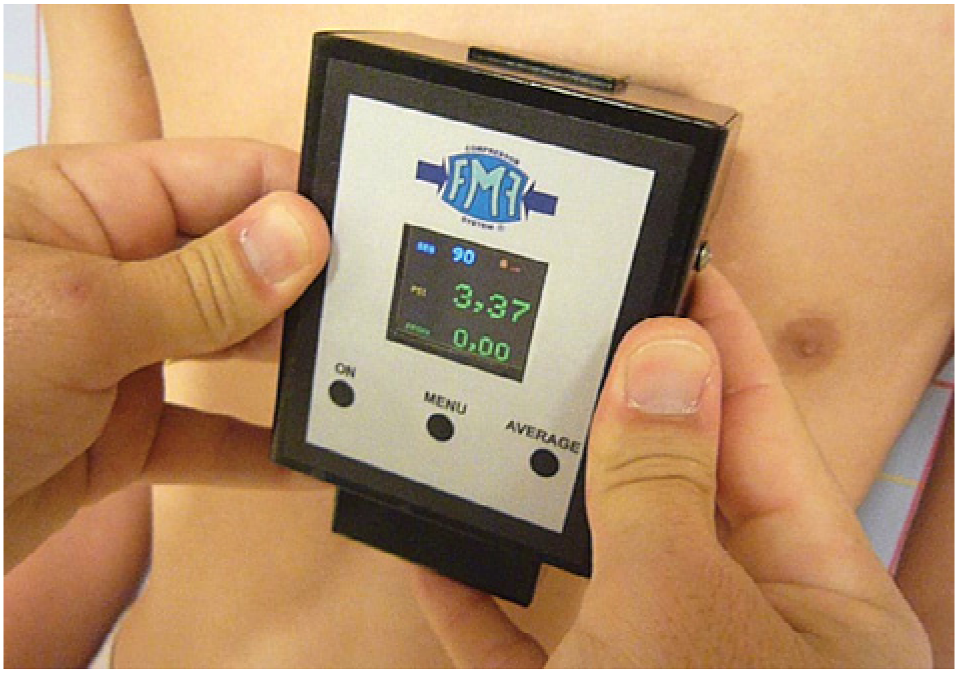
Patient selection and workup

STAGE 1: PATIENT EVALUATION: MEDICAL HISTORY, MEASUREMENTS, AND REQUEST OF THE FMF DCS

The first step in the patient selection process is to distinguish patients who are good candidates for the FMF DCS from those who should be treated surgically.

Specific signs and symptoms possibly associated to pectus carinatum need to be addressed; also, psychological and social impact must be evaluated. The physician will determine if the pectus carinatum is of the chondrogladiolar or chondomanubrial variant. Assessment of symmetry, rigidity of the chest wall, and pain is also mandatory.

The second step is to measure the baseline PC, as shown in **Figure 20.22**. To do this, the PMD will be used. The “AVERAGE” button of the device must be pressed so that it measures the average PC, taking into consideration its variation due to the patient’s respiratory movements.



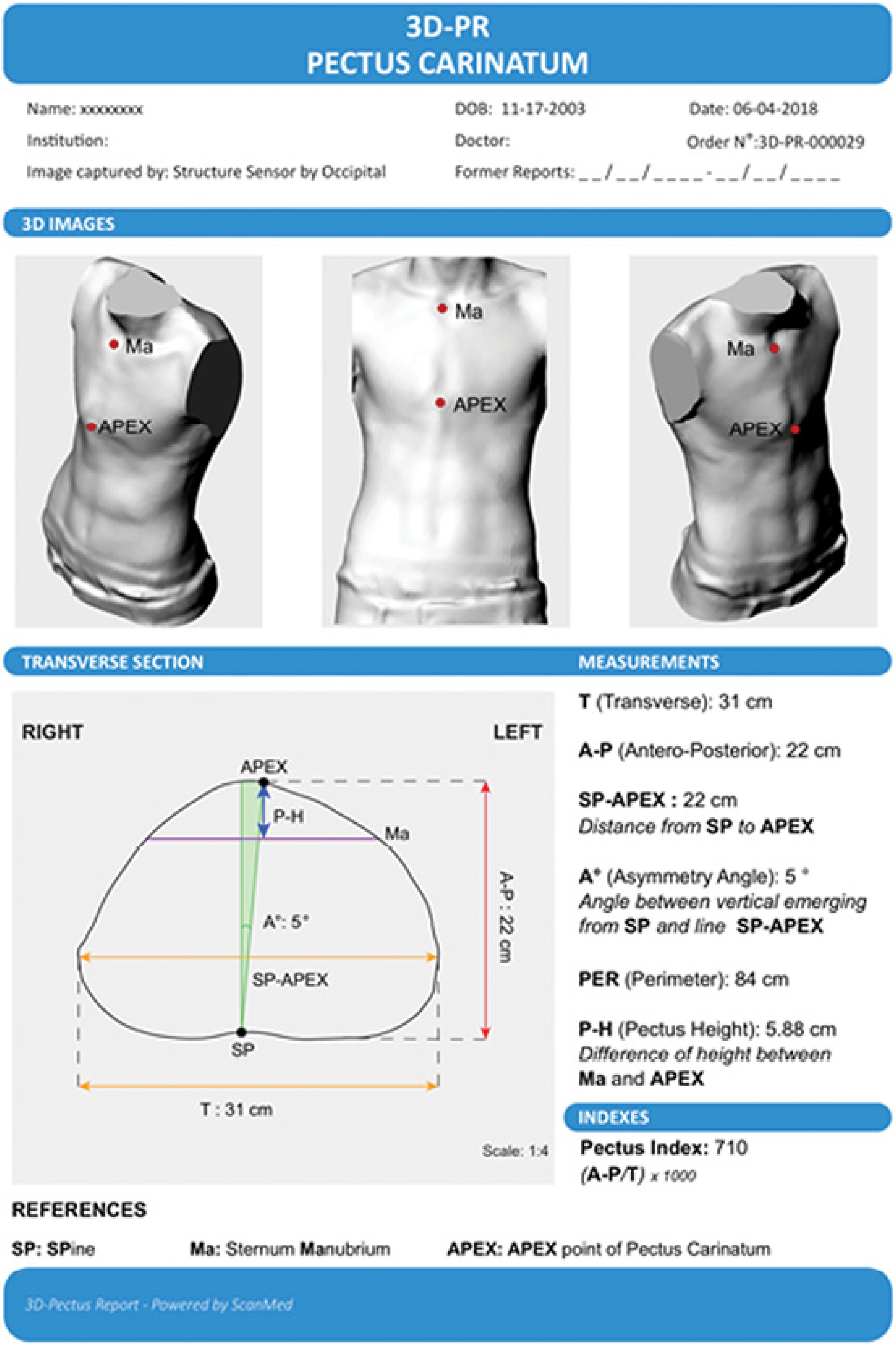
20.22 Measurement of the pressure of correction. The PMD is applied over the skin at the level of the defect and pressure is applied until a visual correction of the defect is achieved. Pressure is measured in pounds per square inch (PSI).

If the pectus carinatum is of chondrogladiolar type and the PC is <14 PSI, a conservative, non-surgical approach should be implemented.

In order to manufacture the custom-fitted brace, specific measurements of the thorax are needed:

- Thoracic perimeter
- Transverse diameter of the chest
- Anterior–posterior diameter of the chest
- Pectus carinatum elevation

In certain institutions this manual measuring method has recently been replaced by a 3D external scanner processed with specific software that will provide detailed measurements of the patient's chest (**Figure 20.23**).



3D-PR
PECTUS CARINATUM

Name: xxxxxxxx	DOB: 11-17-2003	Date: 06-04-2018
Institution:	Doctor:	Order N°:3D-PR-000029
Image captured by: Structure Sensor by Occipital	Former Reports: __/__/____-__/__/____	

3D IMAGES

Ma
APEX
Ma
APEX
Ma
APEX

TRANSVERSE SECTION

RIGHT
LEFT
APEX
P-H
Ma
A-P : 22 cm
A°: 5°
SP-APEX
SP
T : 31 cm
Scale: 1:4

MEASUREMENTS

T (Transverse): 31 cm

A-P (Antero-Posterior): 22 cm

SP-APEX : 22 cm
Distance from ***SP*** *to* ***APEX***

A° (Asymmetry Angle): 5 °
Angle between vertical emerging from ***SP*** *and line* ***SP-APEX***

PER (Perimeter): 84 cm

P-H (Pectus Height): 5.88 cm
Difference of height between
Ma and **APEX**

INDEXES

Pectus Index: 710
(A-P/T) *x 1000*

REFERENCES

SP: SPine　　**Ma:** Sternum **Ma**nubrium　　**APEX: APEX** point of Pectus Carinatum

3D-Pectus Report - Powered by ScanMed

20.23 3D pectus carinatum report as obtained after processing the stereolithography (STL) image obtained with an external 3D scanner. Note that the report provides all the measurements needed for the manufacturing of the custom-made brace and at the same time provides different pectus carinatum indexes for an objective evaluation of the defect.

Patient identification details, PC, thoracic measurements, and position of the pectus carinatum (left, right, center in **Figure 20.23**) are registered in the order form. The size of the compression plate must also be indicated, and, although most patients will do well with the standard size, five other sizes are available, as well as a hollow plate for patients with very sharp-edged pectus carinatum. The number of pads can also range from 1 to 3, depending on the size of the breasts, to maximize comfort in females. A replacement kit briefcase provides the most frequently needed accessories to be used at the pectus carinatum outpatient clinic (**Figure 20.24**).

PRESSURE HOLES
FMI
(a)
(b)

20.24a,b (a) Contoured pad intended for the treatment of "sharp" deformities where the skin may be damaged if direct compression if applied; (b) replacement kit briefcase with the frequently requested elements needed in a pectus carinatum outpatient clinic.

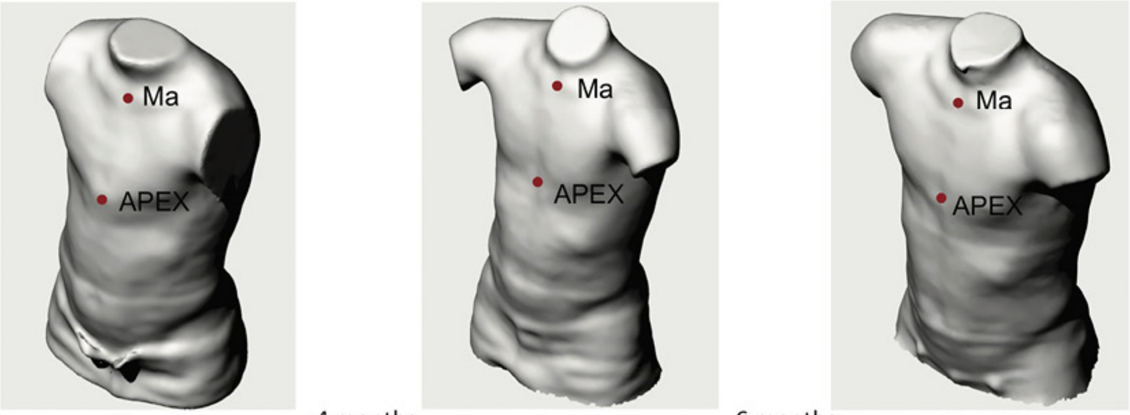
Usually, pictures are taken with the patient's consent, to keep a record of the baseline status and evaluate progress at subsequent visits. In our experience, being able to review the changes with the patient enhances motivation and compliance. A set of six pictures is enough: Left and right 3/4 profile and full profile, front view of the thorax, and front view of the thorax from above. Chest X-ray, CT scan and chest MRI are not routinely indicated. If the patient presents with an atypical or stiff pectus carinatum or severe pain, or belongs to a family that feels insecure about the diagnosis and proposed management, further investigation may be recommended. Although uncommon, if cardiopulmonary symptoms are present, pulmonary function tests and an echocardiogram are a must.

The 3D external scan used for measurement will also deliver digital information that, when processed by a specific software, will provide a pectus carinatum 3D report and carinatum indexes that will be used in further comparative reports to objectively evaluate the evolution of the treatment (**Figure 20.25**).

3D-PR
PECTUS CARINATUM - FOLLOW-UP

Name: xxxxxxxx
DOB: 11-17-2003
Date: 06-04-2018
Institution:
Doctor:
Order N°:3D-PR-000029
Image captured by: Structure Sensor by Occipital
Former Reports: 12/01/2016 - 04/21/2017 - 10/06/2017
Treatement: FMF Dynamic Compressor System

3D IMAGES


Ma
APEX
Ma
APEX
Ma
APEX

Scan 1
12/01/2016

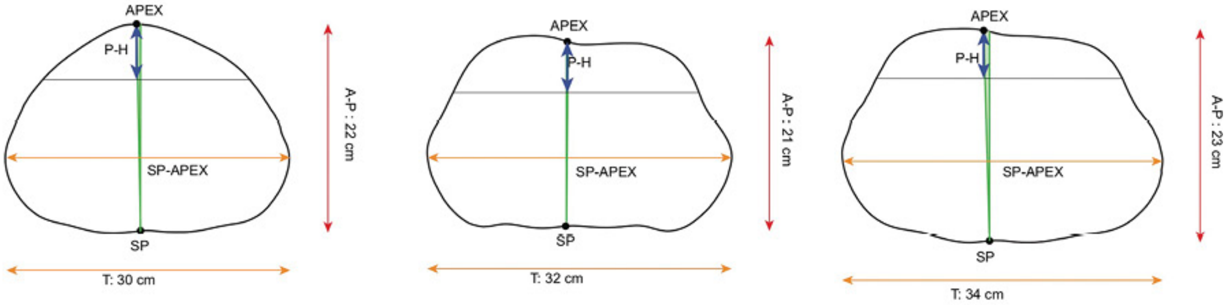
4 months

Scan 2
04/21/2017

6 months

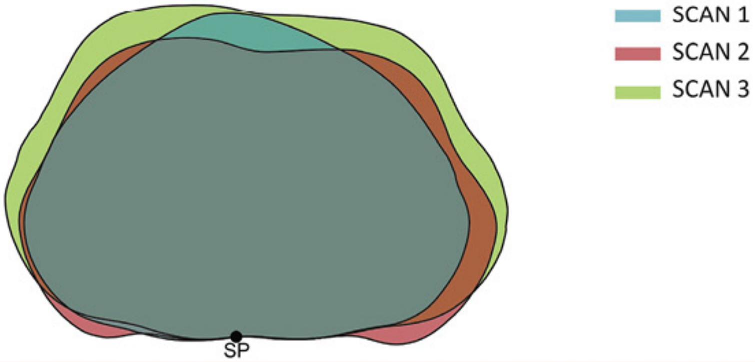
Scan 3
10/06/2017

INDIVIDUAL TRANSVERSE SECTIONS


APEX
P-H
SP-APEX
SP
T: 30 cm
A-P : 22 cm
APEX
P-H
SP-APEX
SP
T: 32 cm
A-P : 21 cm
APEX
P-H
SP-APEX
SP
T: 34 cm
A-P : 23 cm

SCAN 1 SCAN 2 SCAN 3

TRANSVERSE SECTIONS - STACKED


SCAN 1
SCAN 2
SCAN 3
SP

3D-Pectus Report - Powered by ScanMed

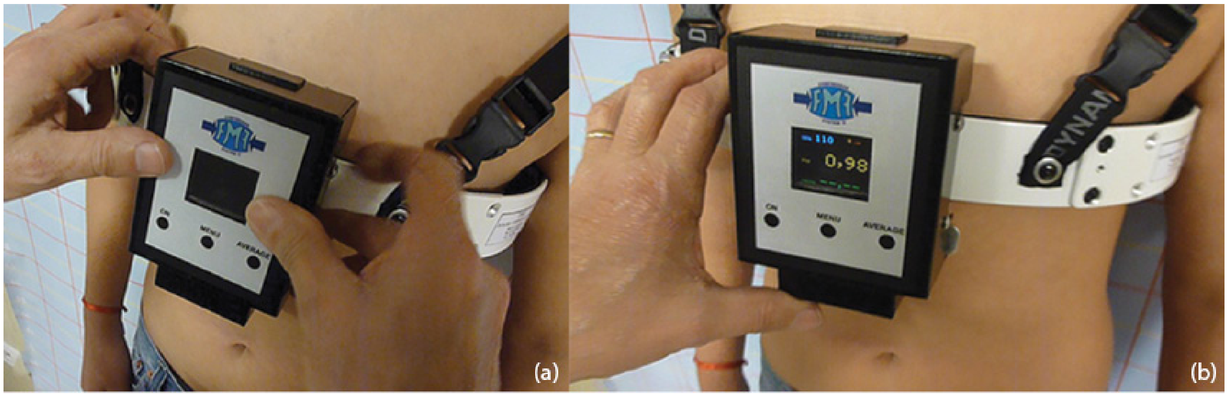
Page 1/2

20.25 3D follow-up pectus carinatum report. This document combines the digital information of three consecutive individual reports and compares them for a precise and objective evaluation of the treatment.

STAGE 2: INITIAL FITTING OF THE BRACE AND PATIENT TRAINING

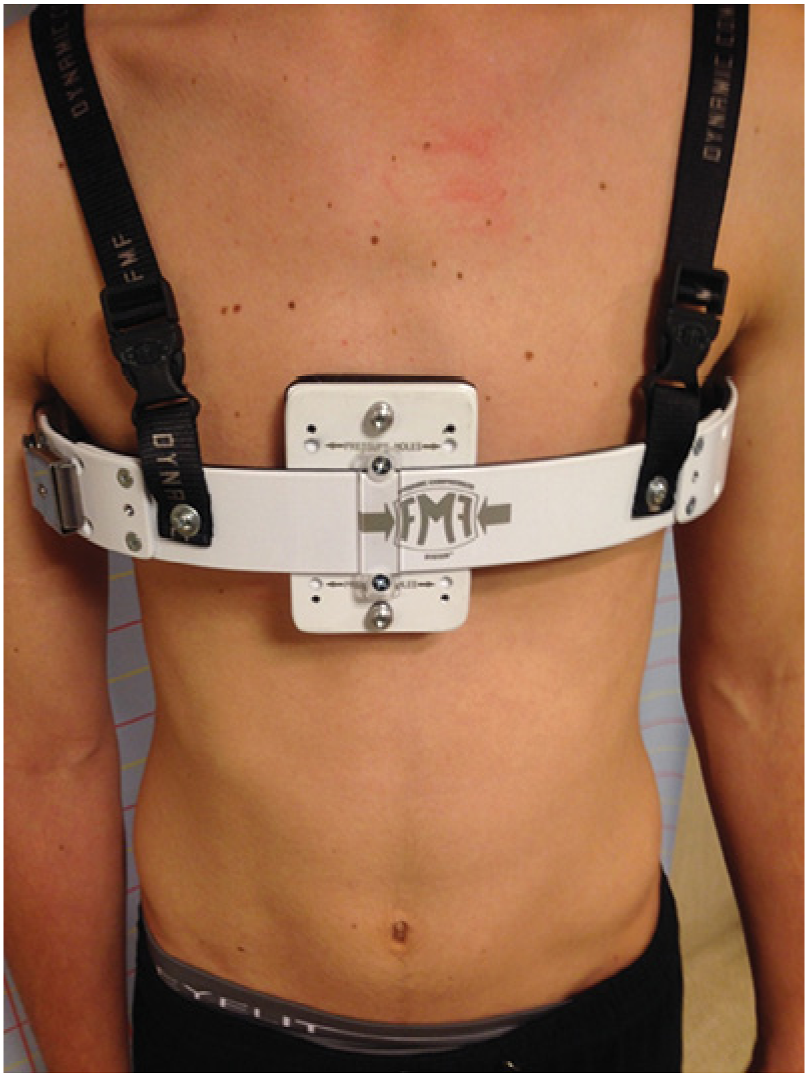
The FMF DCS is received and assembled. The PC is measured again prior to fitting the brace.

Once the brace is fitted, the PT needs to be adjusted, never higher than 2.5 PSI. If the PT is higher than this, the brace will be loosened by using the different adjustment positions (**Figure 20.26a and b**) and the PT will be measured again until satisfactory values are obtained.



20.26a,b (a) Docking the PMD into position; (b) measurement of the pressure of treatment.

The patient is trained so that they can put on and take off the brace by themselves. They are then requested to wear it for 30 minutes and return to the physician's office for evaluation of tolerance of the device. If the skin is too sensitive or the patient refers pain or discomfort to breathe, the PT will be reduced, until the device is well tolerated (**Figure 20.27**).



20.27 FMF DCS in place. Note the available room on each side of the brace, needed for lateral expansion of the chest while the defect is corrected.

Initially, patients will be classified into one of four groups following the treatment guidelines shown in Table 20.1.

Table 20.1 **Recommended use of pressure measured dynamic compression**

	Group 1	Group 2	Group 3	Group 4
PC*	1–4	4–6	6–8	>8
PT**	2.5	2	1.5	1
Use (hours/day)	24	12–24	6–12 (day or night)	3–6 (day or night)
Approximate duration of treatment (months)	2	4	8	12–24

Abbreviations: *PC = Pressure of correction; ** PT = pressure of treatment. Both pressures are measured in PSI.

Notes: Patients are divided into four groups depending on the pressure of correction, which indirectly assesses chest wall elasticity.

Group 1 and 2 patients are instructed to wear the FMF DCS every day, overnight and as much as possible during the day, depending on their activities. They are only allowed to remove the device during sports and while having a shower or bath. We ensure that they understand that the more they wear the device, the faster they will correct the defect.

Patients belonging to Groups 3 and 4 will begin to wear the system for fewer hours per day, at lower PTs, to indirectly increase the flexibility of the thoracic cage, enhance compliance, and prevent skin lesions. We are careful to advise these patients about the complications of overusing the brace and the need to follow medical indications strictly.

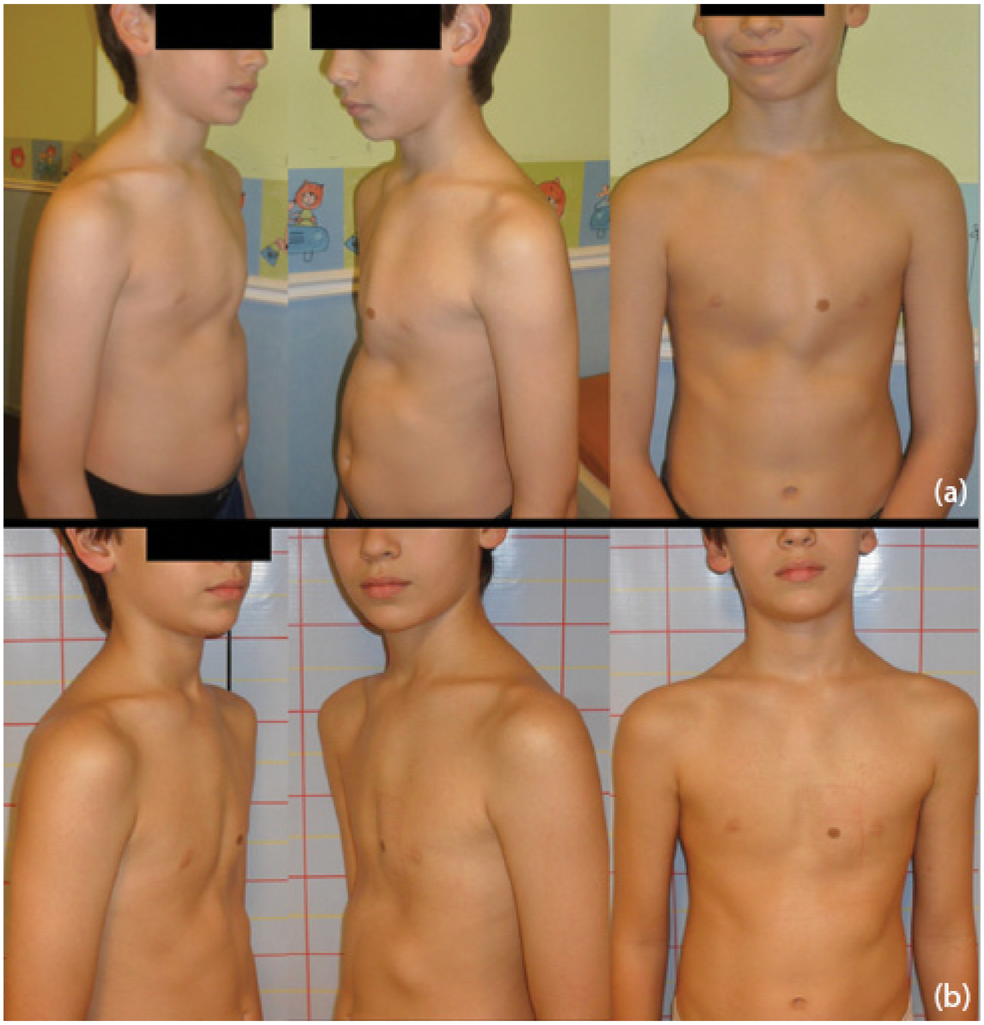
The PC and the duration of treatment results are very useful in helping patients understand what to expect throughout the treatment stages.

STAGE 3: CORRECTION PHASE

The patient has to return for follow-up monthly. During each visit, the PT is measured with the brace setting as it is, and the skin status is evaluated, together with the habits of use of the brace and the practice of complementary activities. The PC is also measured, and the brace is adjusted to set a new PT and usage pattern as described in Table 20.1.

Once the patient's PC lowers to the PC range of the previous group, they are reclassified, and time of usage is adjusted accordingly.

The correction phase ends when the pressure of correction is less than 1 or when the interdisciplinary team, patient, and/or family agree that the deformity has been fully repaired (Figure 20.28).



20.28a,b (a) Patient with a severe chondrogladiolar right-sided pectus carinatum before treatment. (b) Results obtained after 4 months of intensive treatment. The correction phase is concluded and at this stage the patient is ready to start the weaning phase.

STAGE 4: WEANING PHASE

Once the defect is reverted, the FMF DCS is gradually withdrawn to avoid eventual partial recurrences. Pectus carinatum may return mildly, in approximately 10% of cured patients, particularly if they have been treated before pubertal growth spurts or in cases where they have cured very rapidly.

During the weaning phase, patients wear the brace as a "retainer" during the day or overnight (they generally prefer the latter), every day for the first month, every 2 days for the second month, and every 3–4 days for the next few months (range: 2–6 months).

The faster the patient gets cured, the longer the weaning phase should be. PT will remain fixed during the post-correction period. The weaning period is not contemplated in the calculation of the duration of treatment.

STAGE 5: FOLLOW-UP PHASE

Provisory treatment interruption is indicated when the weaning period ends. Patients are controlled every 6 months until they are 18 years old. In adults, treatment finishes when the defect is corrected. Nonetheless, they are all instructed to return if any partial recurrence is observed so that it can be corrected with a few more months of treatment with the brace.

For this phase, the objective evaluation of the patient's evolution is performed by means of periodic 3D external wscanners. A detailed comparative 3D pectus report provides all the necessary information as well as graphics and tables with different pectus carinatum external indexes (see **Figure 20.25**).

Results

Since its first publication in 2008, the FMF DCS has become the standard of care for many centers dedicated to the treatment of chest wall deformities around the world.

A recent systematic review that included 8 out of 14 studies collected a total of 1185 patients treated between 2008 and 2018. The median age was 14 years (range 2–28 years) and 87% were male. The mean study follow-up period was 16 months; 44% of patients were still under treatment, 29% of patients successfully completed treatment, 6% dropped out, and 21% were lost to follow-up. Dropout was mainly caused by social discomfort (7.2%) and failure of treatment (5.8%). Complications were infrequent. Mild chest discomfort or tightness was reported in 12% and skin lesions occurred in 5.1%. The overall recurrence rate was 2.6%. The authors of this systematic review concluded that measured dynamic compression appears to be a safe, non-invasive, and efficient treatment to correct pectus carinatum in patients with a non-rigid thorax. Patients experienced less discomfort, which in turn resulted in better compliance. However, accurate selection of patients based on age, pressure of initial correction, and motivation is important and an objective scoring system to assess the esthetic and long-term physical and psychological results of the treatment is needed.

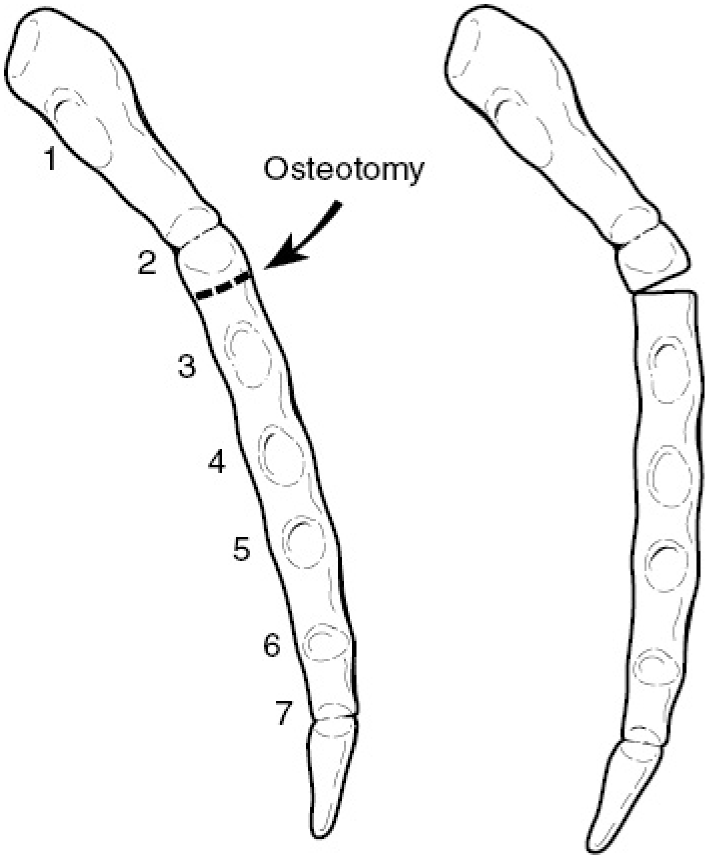
OPERATIONS

Pectus carinatum presents in three primary configurations. The most common is protrusion of the body of the sternum (gladiolous) and costal cartilages and is termed the "chondrogladiolar protrusion." This protrusion can be asymmetric, producing a keel-like protrusion along one side of the sternum. As mentioned previously, some children can have a "mixed'" deformity with ipsilateral carinate and contralateral depression components. The sternum is often rotated to one side in these cases. The rarest configuration is the chondromanubrial protrusion, also referred to as a "pouter pigeon breast." In this deformity, the manubrium and the first and second costal cartilages protrude, and there is relative depression of the body of the sternum.

Open repair

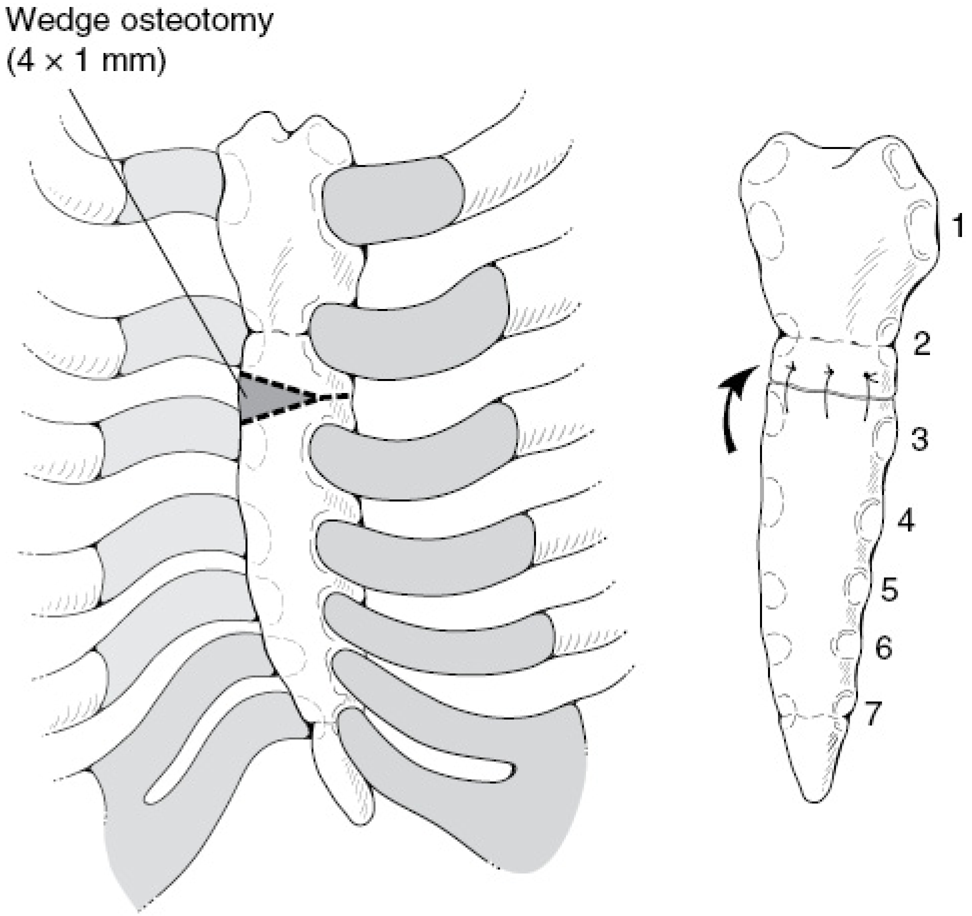
The exposure of the sternum and costal cartilages is identical with that of the open repair for pectus excavatum. The protruding costal cartilages are resected with attention to maximum preservation of the perichondrial sheaths.

A single or double transverse osteotomy with fracture of the posterior plate after resection of the costal cartilages allows posterior displacement of the sternum to an orthotopic position (**Figure 20.29**).



20.29

The mixed pectus deformity is corrected by full and symmetric resection of the involved costal cartilages, followed by a transverse offset (0–10-degree wedge-shaped sternal osteotomy) (**Figure 20.30**). Closure of this defect achieves both anterior displacement and rotation of the sternum. In some patients use of plates and screws to stabilize the osteotomy is advantageous.



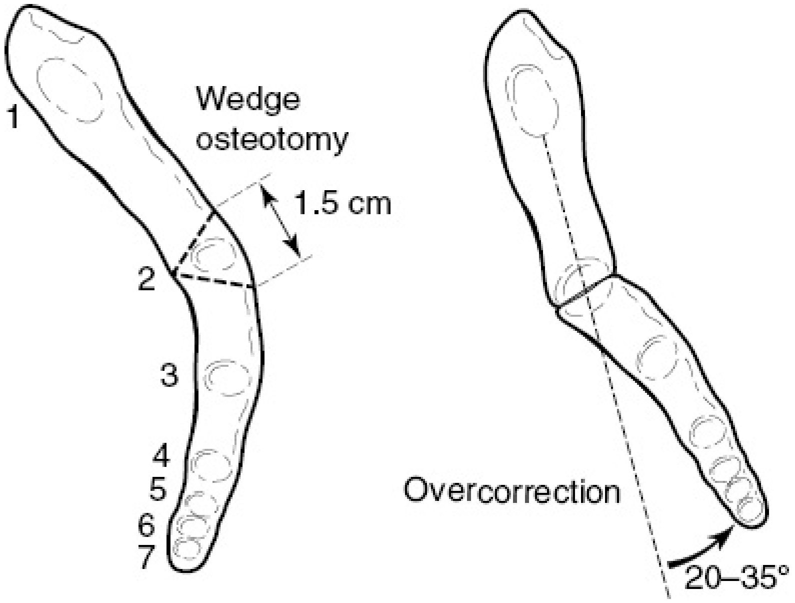
Mixed deformity (anterior view) 9.2%

20.30

OUTCOME

Generally, excellent results are achieved with repair of pectus carinatum. Repair at a young age may result in development of a protrusion of cartilages which have not been resected, or in cases of a unilateral abnormality, contralateral protrusion may occur. For this reason, correction of pectus carinatum is generally deferred until children have completed most of their pubertal growth.

In the chondromanubrial deformity, both the protrusion of the manubrium and depression of the body of the sternum must be corrected. A broad, wedge-shaped sternal osteotomy is placed through the anterior cortex of the obliterated sternomanubrial junction. Closure of the osteotomy after fracture of the posterior cortex achieves posterior displacement of the superior portion of the sternum, which is attached only by its junction with the first rib. The lower portion of the sternum is overcorrected 20–35 degrees and is secured in position by strut, suture, or plate and screw fixation, correcting both components of the abnormality (**Figure 20.31**).

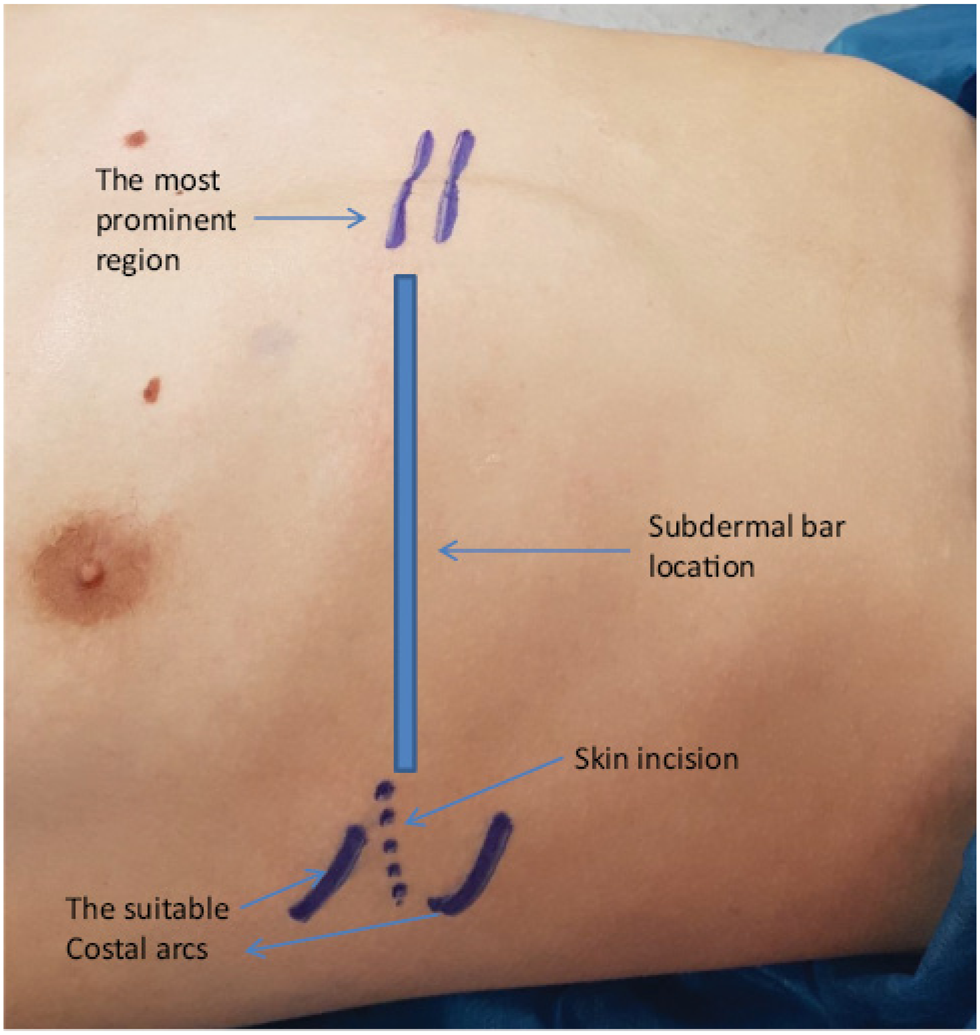


20.31

Minimally invasive repair: Abramson procedure

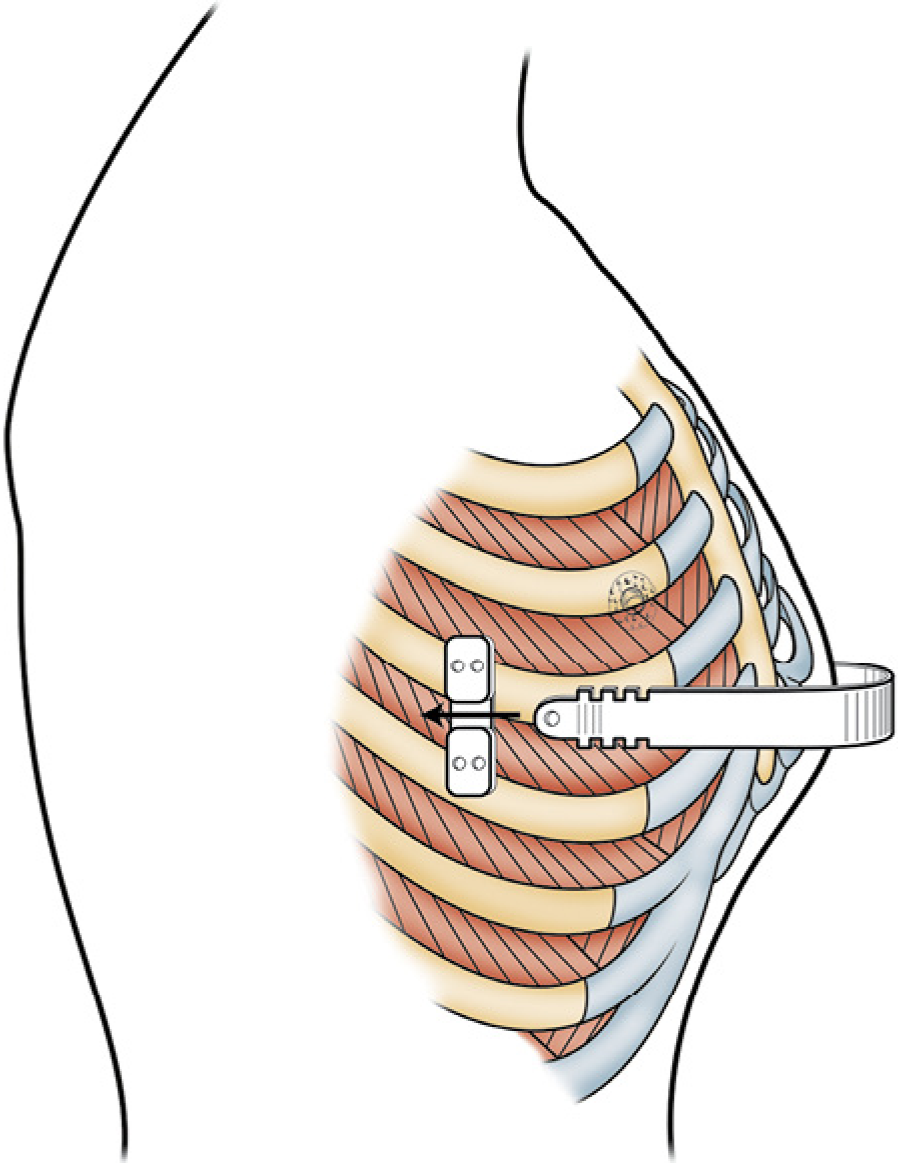
The minimally invasive operation developed by Abramson is most successful in patients with a flexible chest. Since most such patients are candidates for brace treatment, the operation is employed for flexible chest patients who cannot or will not try brace treatment, or who fail brace treatment due to failure to wear the brace as instructed.

For the Abramson operation, the patient is positioned supine with both arms abducted about 75 degrees at the shoulders, and with the elbows slightly flexed, with rolled-up foam under the wrists to achieve a relaxed upper extremity. The most prominent portion of the carinatum is identified, and a bar is chosen to compress the chest at that site. The skin is marked there (**Figure 20.32**).


The most prominent region
Subdermal bar location
Skin incision
The suitable Costal arcs

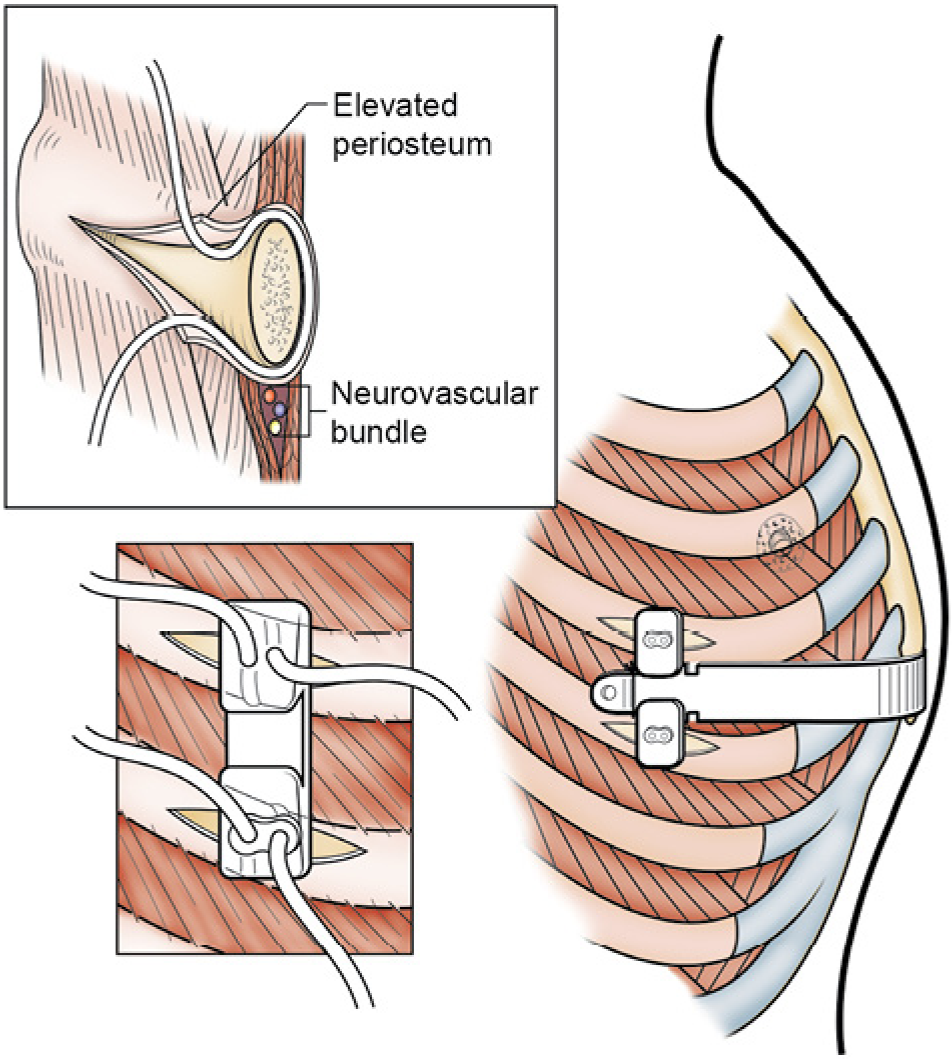
20.32 Marking the patient.

The bar length should allow the bar to extend from one mid-axillary line, across the chest in its corrected position, to the other mid-axillary line (**Figure 20.33**). A flexible metal template in the shape of the bar is applied to the chest as an assistant compresses the protrusion (**Figure 20.33**). The template can be used to guide bending the bar.



20.33 Bending the bar to the desired chest wall shape.

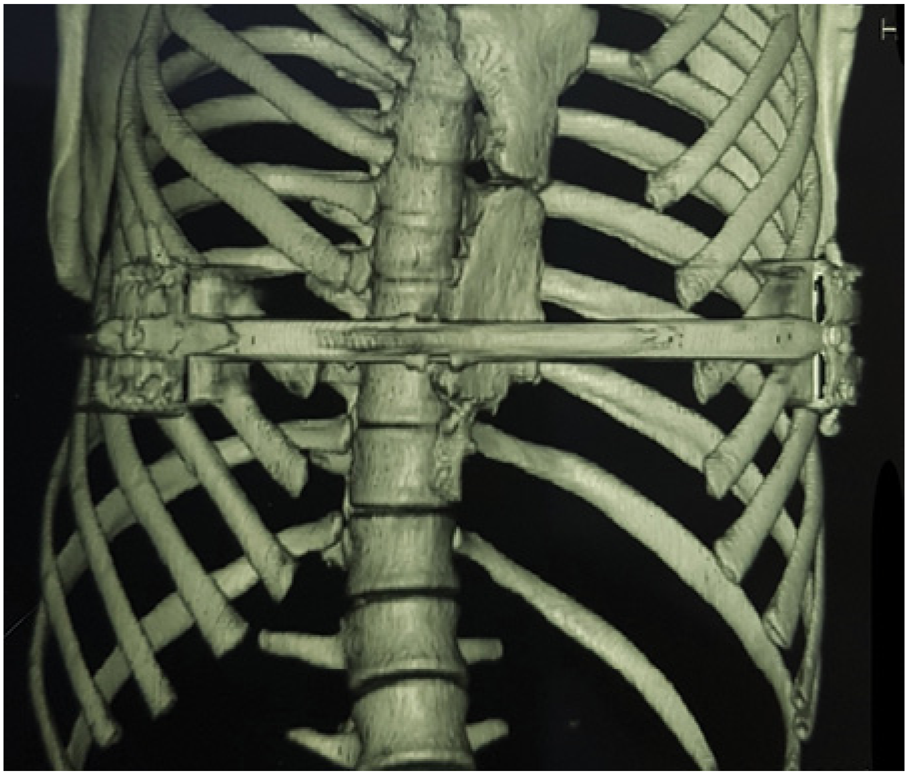
The bar is bent to conform to the desired chest shape at that axial level. It is important that the bar rests on the sternum itself. Ribs are chosen for attachment of the bar to the chest subperichondrially at approximately the mid-axillary line (**Figure 20.34**). In female patients, the incision is placed where the breast meets the chest wall (laterally) in a gentle curve. In males, horizontal incisions are placed directly over the ribs chosen to anchor the bar.



20.34 Attaching the stabilizer to the ribs subperichondrially.

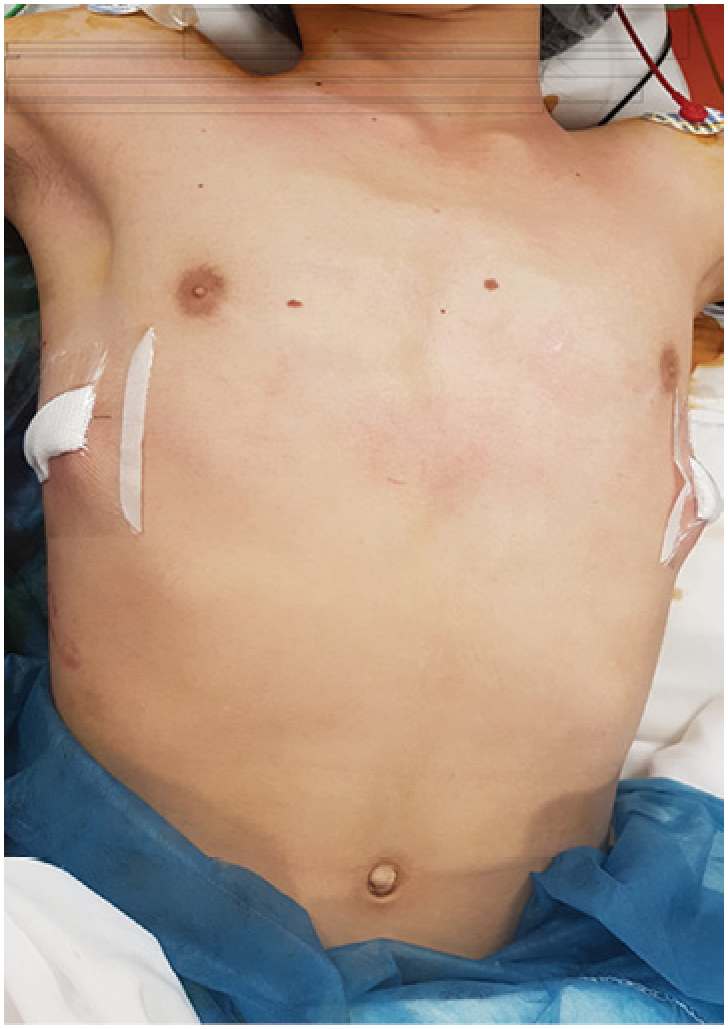
Lateral thoracic incisions are made, and the ribs are dissected free. A subperiosteal tunnel is created around each of four ribs. Plastic tubes (such as are used for Rammel tourniquets) are passed through these, and steel wires or cables are passed through the tubing around the ribs. Pockets suitable for accommodating the fixation plates are created submuscularly. The wires or cables are threaded through the holes in the plates.

The path for the bar is created posterior to the pectoralis major or subcutaneously depending on the location of the protrusion. If the protrusion is inferior to the origin of the pectoralis major, the tunnel will be subcutaneous. This can be accomplished with long scissors, a Crawford clamp, a Zimmer-Biomet Nuss bar introducer, or other device. The instrument is passed through the pectoralis major medially, across the sternal protrusion, and then brought subpectorally on the other side. A 28 Fr chest tube is then guided back across the tunnel and is used to guide the bar into position. The chest is then again compressed while the bar is introduced into the stabilizer plates, and it is secured to them by screws or wires (depending on available equipment). The wires or cables around the ribs are then tightened. The pressure on the chest is slowly changed to allow the desired thoracic contour to be reached (**Figure 20.35**).

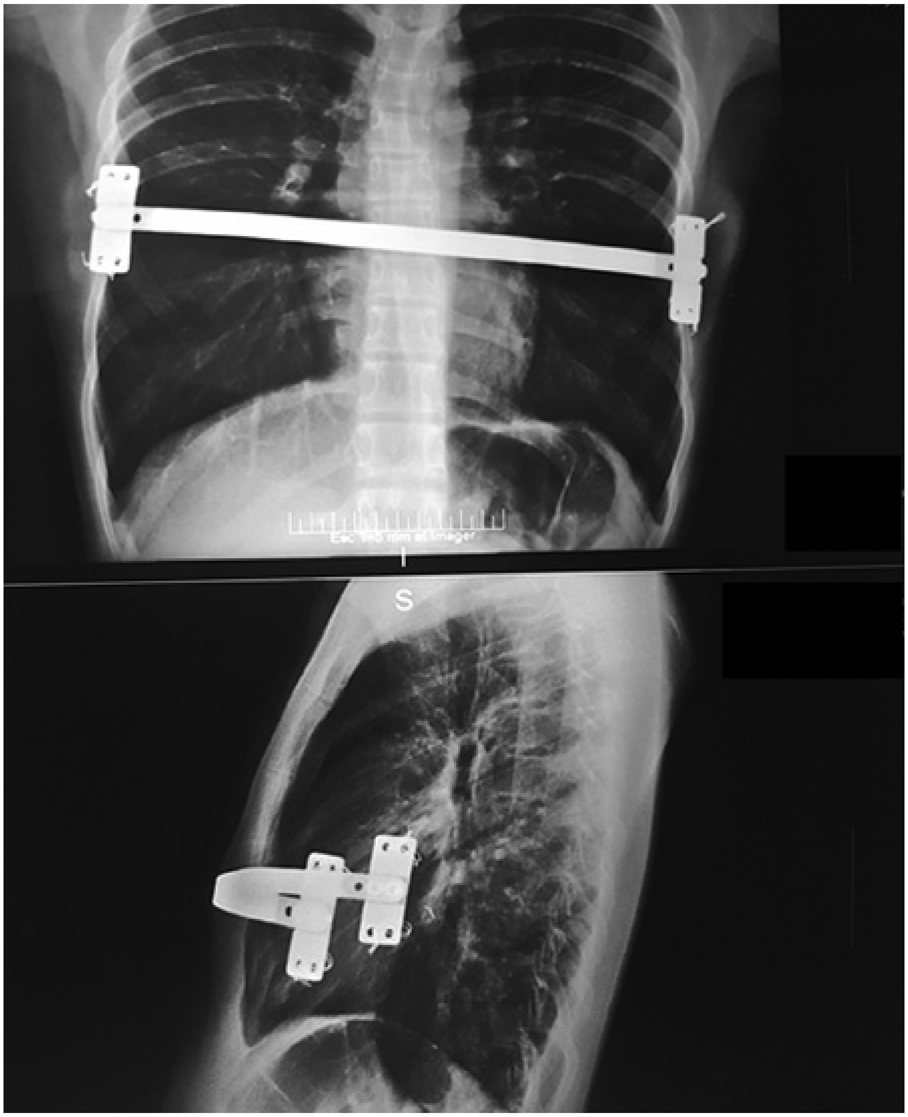


20.35 Three-dimensional image of the bar in place.

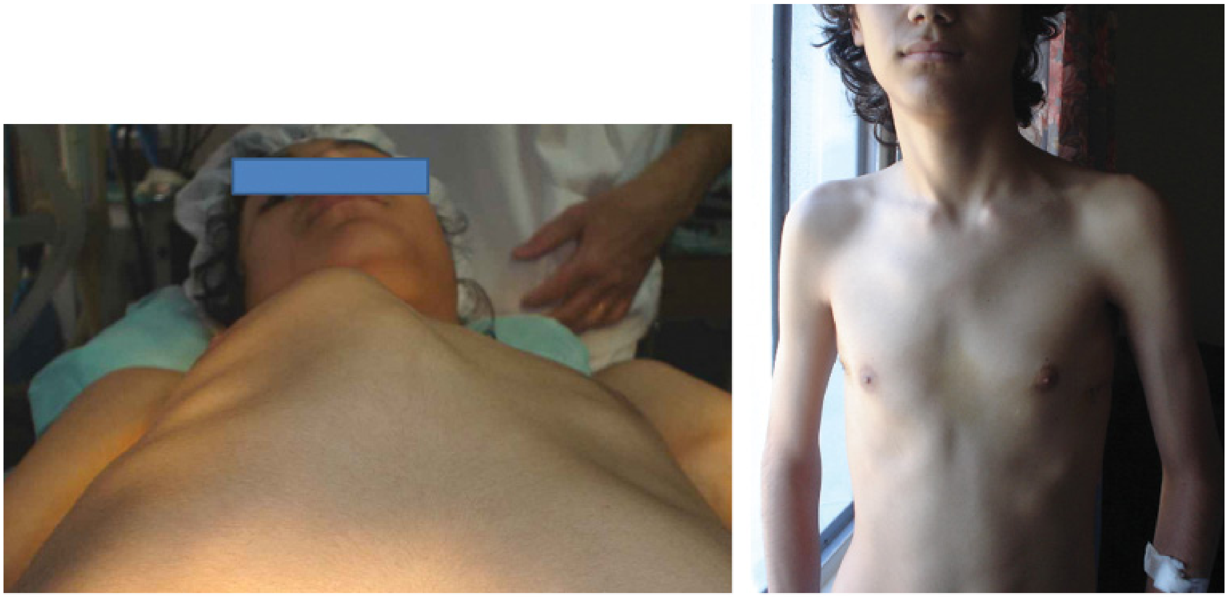
The wound is then filled with saline while positive pressure is applied to the endotracheal tube to look for pneumothorax. Layered closure is followed by chest X-ray (**Figures 20.36– 20.38a and b**).



20.36 Postoperative chest appearance.



20.37 Postoperative chest X-ray.


[redacted]

(a) (b)

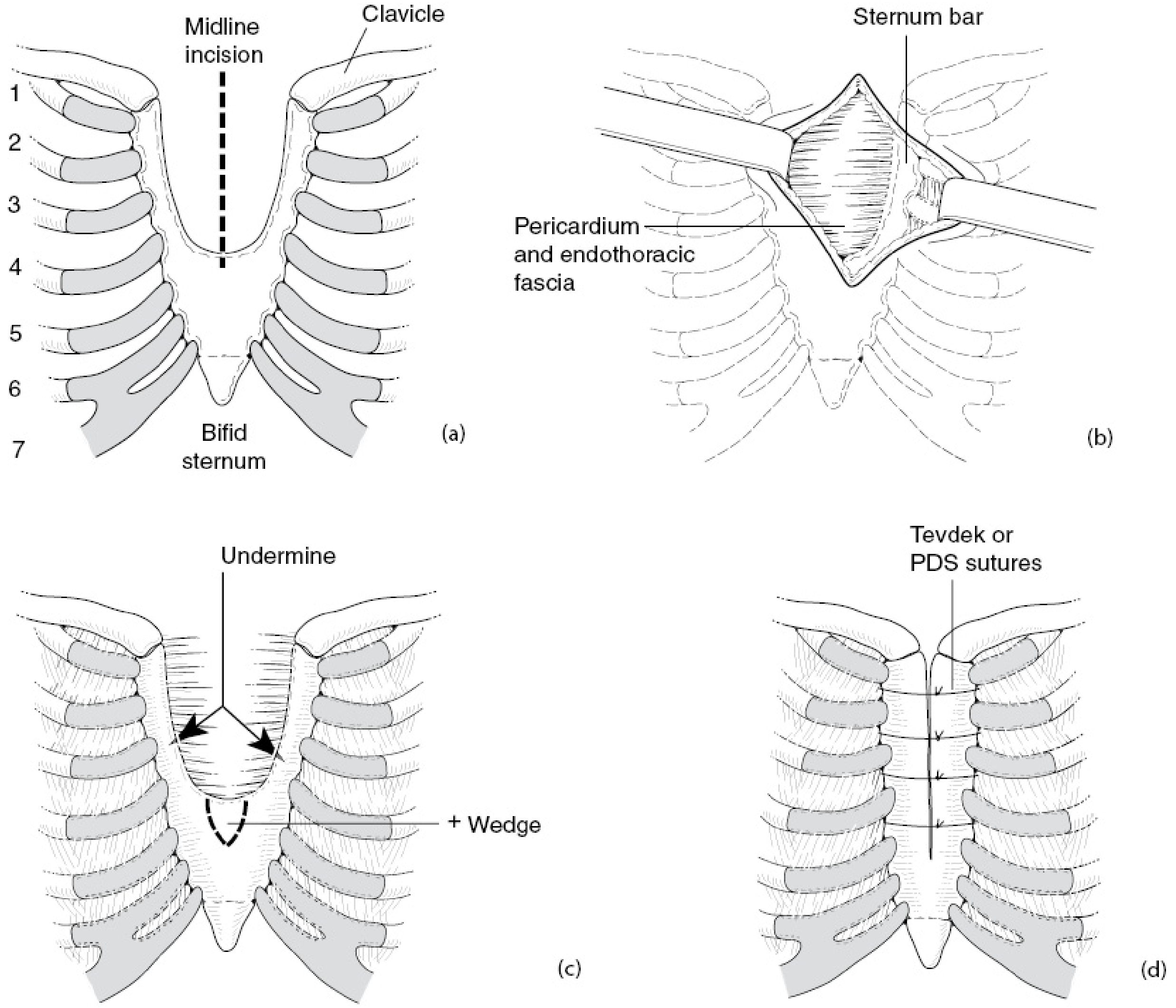
20.38a,b Severe asymmetric pectus carinatum (a) before repair and (b) 24 hours after minimally invasive correction.

STERNAL CLEFT

The sternal cleft deformity is the least severe of the sternal abnormalities, which also include thoracic and thoracoabdominal ectopia cordis. A sternal cleft results from incomplete fusion of the sternal bars in the fetus. The heart is in an orthotopic position. While intrinsic cardiac anomalies are common in thoracoabdominal and thoracic ectopia cordis, they are rare in infants with a sternal cleft. Repair of the sternal cleft relieves the paradoxical motion often seen in this anomaly. The sternal cleft may be complete, but in most cases the inferior base of the sternum is fused. Repair is best achieved in the newborn period or within the first several months of life when the chest wall is most flexible and primary repair is tolerated by the infant.

OPERATION

Repair of the bifid sternum is best performed through a longitudinal incision extending the length of the defect (**Figure 20.39a**). Directly beneath the subcutaneous tissues, the sternal bars are encountered. The pectoral muscles insert lateral to the bars (**Figure 20.39b**). The endothoracic fascia is mobilized off the sternal bars posteriorly with blunt dissection to allow safe placement of the sutures (**Figure 20.39c**). In many cases, excision of a wedge of cartilage from the most inferior portion of the defect will facilitate approximation of the two sternal halves during suture closure. Closure of the defect is achieved with 2/0 Tevdek or PDS sutures (**Figure 20.39d**).


Midline incision
Clavicle
1
2
3
4
5
6
7
Bifid sternum
(a)
Sternum bar
Pericardium and endothoracic fascia
(b)
Undermine
+ Wedge
(c)
Tevdek or PDS sutures
(d)

20.39a–d

More complex repairs are required in older children. These include bilateral oblique incisions through the costal cartilages to increase their length, allowing midline approximation of the sternal halves, or by division of the cartilages, allowing them to be swung medially to cover the defect.

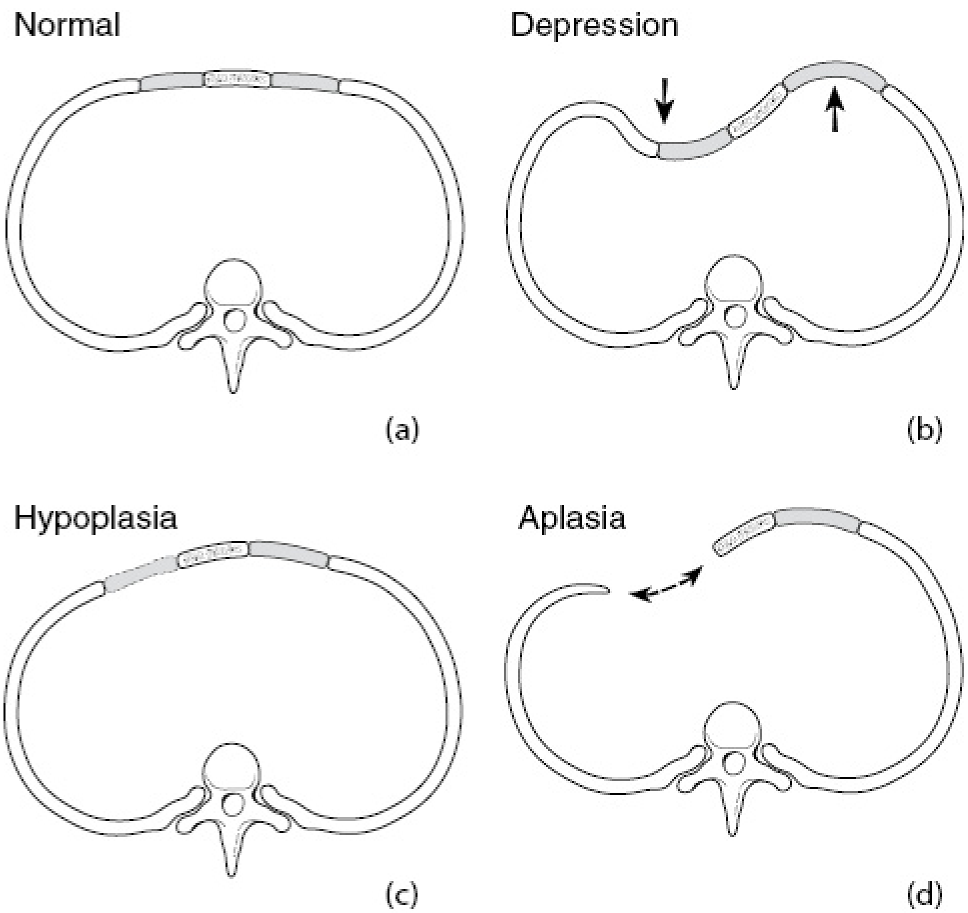
OUTCOME

Repair at an early age is most satisfactory. Closure with cartilage or rib grafts often leads to instability of the chest with progressive growth. Pectus excavatum has been reported to occur years after repair.

POLAND'S SYNDROME

Poland's syndrome is a constellation of anomalies including absence of the pectoralis minor muscle, absence of the costal portion of the pectoralis major muscle, hypoplasia of the breast and nipple or complete absence of the breast and nipple, and brachysyndactyly of the digits. A variable deformity of the chest wall occurs ranging from hypoplasia to a severe excavatum deformity to aplasia of several ribs.

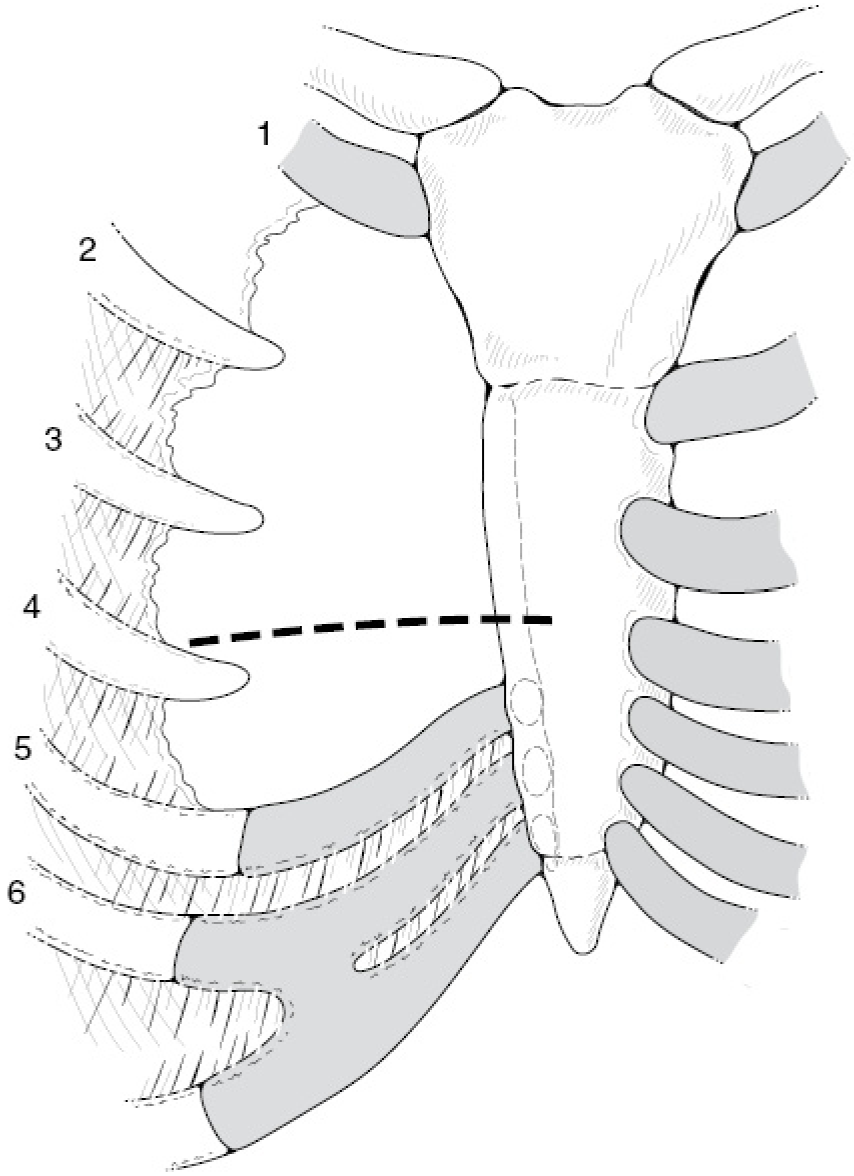
Figures 20.40a–d depict the spectrum of thoracic abnormality seen in Poland's syndrome. Usually, an entirely normal thorax is present, and only the pectoral muscles are absent (**Figure 20.40a**). There may be depression of the involved side of the chest wall, with rotation and often depression of the sternum. A carinate protrusion of the contralateral side is frequently present (**Figure 20.40b**). Hypoplasia of ribs on the involved side but without significant depression may be seen (**Figure 20.40c**). This usually does not require surgical correction. Aplasia of one or more ribs is usually associated with depression of adjacent ribs on the involved side and rotation of the sternum (**Figure 20.40d**).



20.40 a–d

This deformity has its gravest implication in females in whom significant abnormality of the breast is seen. In females, repair of the underlying chest wall depression facilitates correction of the absence or hypoplasia of the breast with a prosthetic implant. Often simultaneous rotation of the latissimus dorsi muscle is performed to create a more natural texture to the breast. In males, repair of the aplastic segment is undertaken in adolescence to avoid impairment of growth resulting from surgery at an early age.

Figure 20.41 is a schematic depiction of a severe deformity with rotation of the sternum and aplasia of three ribs. Ironically, the contralateral side of the chest may have a carinate protrusion which accentuates the depression on the ipsilateral side. The rotation of the latissimus dorsi muscle that is occasionally utilized in males has the potential drawback of decreasing strength of the shoulder.

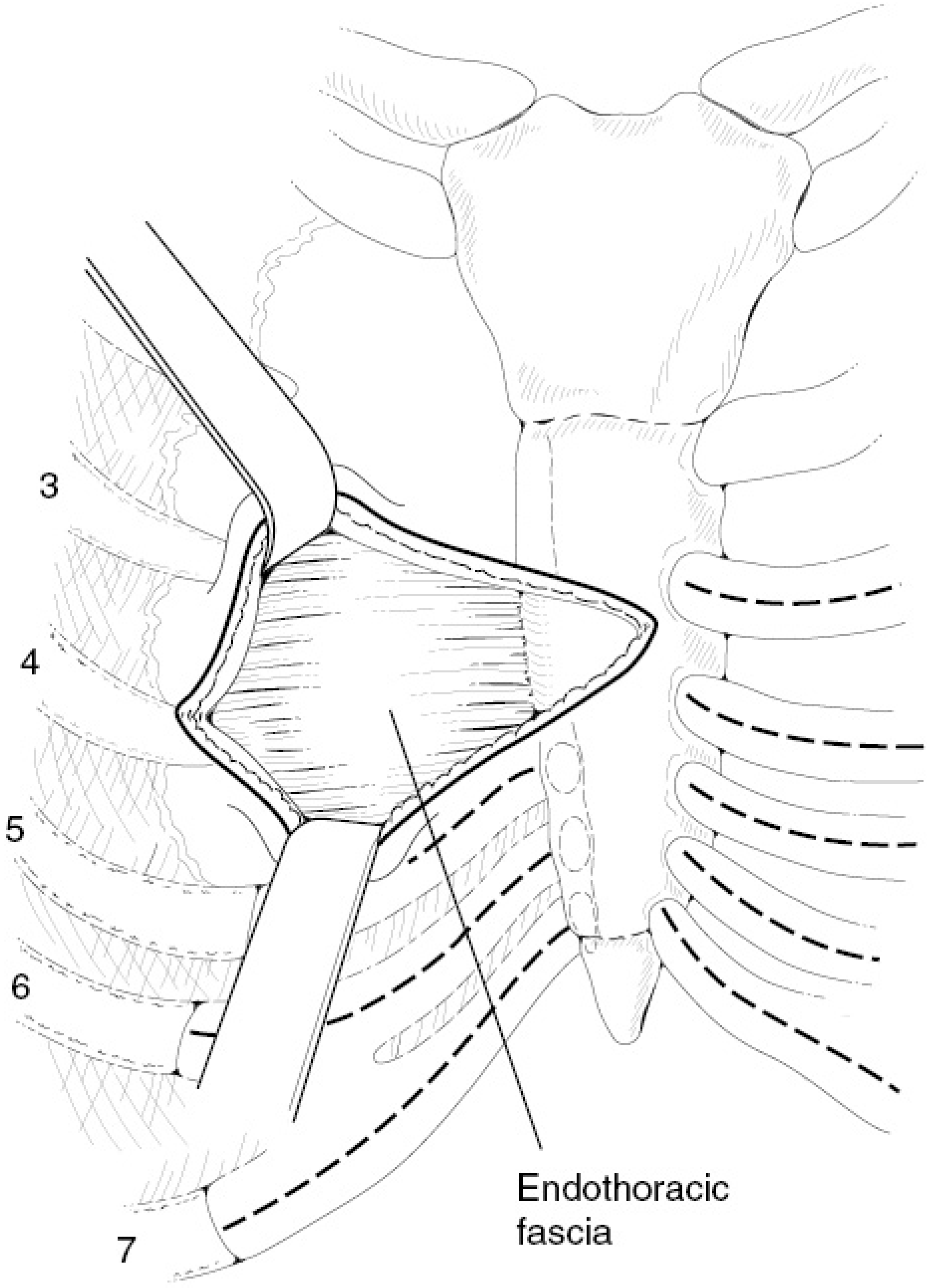


20.41

OPERATION

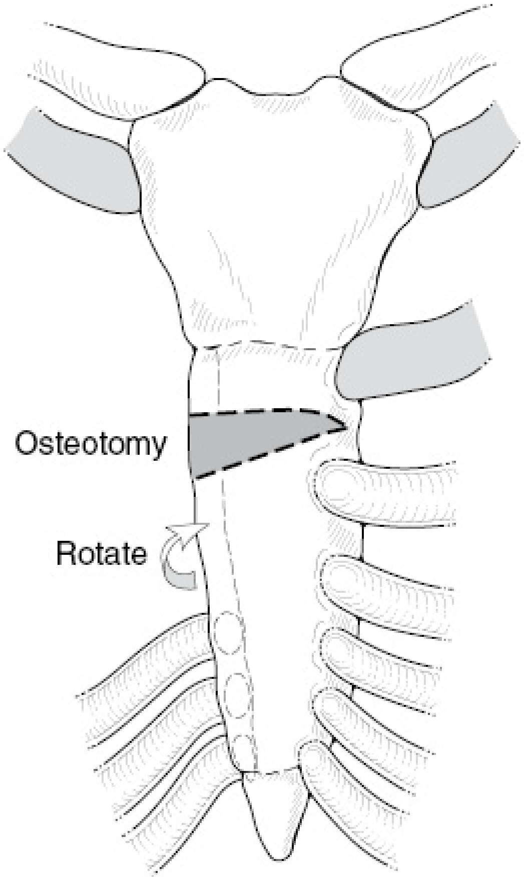
The incision for repair is similar to that in pectus excavatum and carinatum. A transverse incision is placed below the nipples and, in females, in the inframammary crease.

In patients with aplasia of the ribs, the endothoracic fascia is encountered directly below the attenuated subcutaneous tissue and pectoral fascia (**Figure 20.42**). The pectoral muscle flap is elevated on the contralateral side and the pectoral fascia, if present, on the involved side. Subperichondrial resection of the costal cartilages is then carried out, as shown by the bold dashed lines in the figure. Rarely, this must be carried to the level of the second costal cartilages.



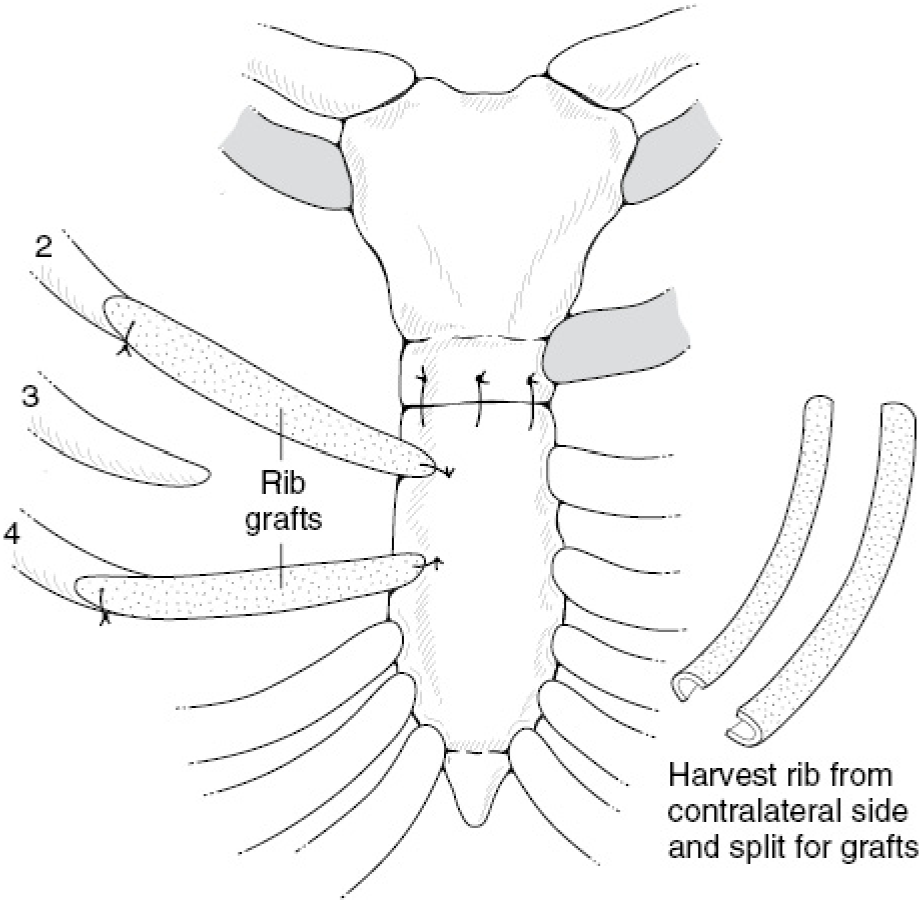
20.42

A transverse, offset, wedge-shaped sternal osteotomy is created below the second costal cartilage (**Figure 20.43**). Closure of this defect with heavy silk sutures or elevation with a retrosternal strut corrects both the posterior displacement and the rotation of the sternum.



20.43

In patients with rib aplasia, split rib grafts are harvested from the contralateral fifth or sixth rib and then secured medially with wire sutures into previously created sternal notches and with wire to the native ribs laterally. Ribs are split as shown in **Figure 20.44** along their short axis to maintain maximum mechanical strength.


2
3
4
Rib grafts
Harvest rib from contralateral side and split for grafts

20.44

OUTCOME

A broad spectrum of chest wall abnormalities is seen in Poland's syndrome, and surgical intervention must be appropriately tailored to the severity of the deformity. Hypoplasia of the chest without a localized posterior depression does not require repair. A significant posterior depression, particularly in females, warrants repair in adolescence.

ACKNOWLEDGMENTS

Figures 20.1–20.5 and 20.8 are from Shamberger RC, Welch KJ. Surgical repair of pectus excavatum. *J Pediatr Surg* 1988; 23: 615, with permission. **Figure 20.4** is adapted from the original figure. **Figures 20.6 and 20.7** are

from Shamberger RC. Chest wall deformities. In: Shields TW (ed.). *General Thoracic Surgery*, 5th ed., 2005, with permission from Lippincott, Williams & Wilkins.

Figures 20.9 and 20.10 are reproduced from Shamberger RC. Depression deformities: Pectus excavatum. In: *Pediatric Surg and Urology: Long-term Outcomes*, 1998, with permission from WB Saunders.

Figures 20.29 and 20.30 are reproduced with permission from Shamberger RC, Welch KJ. Surgical correction of pectus carinatum. *J Pediatr Surg* 1987; 22: 48–53.

Figure 20.31 is reproduced with permission from Shamberger RC, Welch KJ. Surgical correction of chondromanubrial deformity. *J Pediatr Surg* 1988; 23: 319.

Figure 20.39a–d is reproduced with permission from Shamberger RC, Welch KJ. Sternal defects. *Pediatr Surg Int* 1990; 5: 156.

Figures 20.40a–d and 20.44 are reproduced with permission from Shamberger RC, Welch KJ, Upton J III. Surgical treatment of thoracic deformity in Poland's syndrome. *J Pediatr Surg* 1989; 24: 760.

The authors gratefully acknowledge the efforts of Drs Robert Shamberger, Michael Goretsky, and Donald Nuss, authors of this chapter in the previous edition, who supplied figures and text utilized in this work.

FURTHER READING

Abramson H, D'Agostino J, Wuscovi S. A 5-year experience with a minimally invasive technique for pectus carinatum repair. *J Pediatr Surg* 2009; 44: 118–23.

de Beer SA, Blom YE, Lopez M, de Jong JR. Measured dynamic compression for pectus carinatum: A systematic review. *Semin Pediatr Surg* 2018; 27: 175–82.

de Beer SA, Gritter M, de Jong JR, van Heurn ELW. The dynamic compression brace for pectus carinatum: Intermediate results in 286 patients. *Ann Thorac Surg* 2017; 103(6): 1742–9.

Horth L, Stacey MW, Proud VK, et al. Advancing our understanding of the inheritance and transmission of pectus excavatum. *J Pediatr Genet* 2012; 1: 161–73.

Kelly RE Jr, Daniel A. Outcomes, quality of life, and long-term results after pectus repair from around the globe. *Semin Pediatr Surg* 2018; 27: 170–4.

Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. *Ann Surg* 2010; 252: 1072–81.

Kelly RE Jr, Shamberger RC, Mellins RB, et al. Prospective multicenter study of surgical correction of pectus excavatum: Design, perioperative complications, pain, and baseline pulmonary function facilitated by internet-based data collection. *J Am Coll Surg* 2007; 205: 205–16.

Obermeyer RJ, Cohen NS, Jaroszewski DE. The physiologic impact of pectus excavatum repair. *Semin Pediatr Surg* 2018; 27: 127–32.

Obermeyer RJ, Cohen NS, Kelly RE Jr, et al. Nonoperative management of pectus excavatum with vacuum bell therapy: A single center study. *J Pediatr Surg* 2018; 53: 1221–5.

Obermeyer RJ, Gaffar S, Kelly RE Jr, et al. Selective versus routine patch metal allergy testing to select bar material for the Nuss procedure in 932 patients over 10 years. *J Pediatr Surg* 2018; 53: 260–4.

21

Patent ductus arteriosus

AARON R. JENSEN AND JAMES E. STEIN

PRINCIPLES AND JUSTIFICATION

The ductus arteriosus of many preterm infants remains patent at birth. Patency approaches 90% under 28 weeks' gestation, 75% at 28–31 weeks, 45% at 31–33 weeks, and 21% at 34–36 weeks. When correlated with weight, there is 83% patency under 1000 g; this falls to 47% in those weighing 1000–1500 g at birth and 27% in neonates weighing more than 1500 g. While spontaneous closure is directly related to arterial oxygen tension, there are many additional factors that affect functional and anatomic closure, and reopening, in both the premature and full-term infant.

Indications

Historically, it was thought that up to 70% of infants delivered before 28 weeks' gestation will need the ductus closed, medically or surgically. This practice, however, has changed over the past decade with a trend toward observation and allowing spontaneous closure. The use of pharmacologic intervention and surgical ligation has decreased over this time without a noted difference in clinical outcomes. While most of these data are derived from administrative databases and lack granularity to elucidate differences in indication for ligation, an increase in conservative management is likely to continue until more detailed investigation into the true benefit by indication is ascertained.

A patent ductus arteriosus (PDA) results in increased blood flow to the lungs, exposing them to systemic blood pressures. The left-to-right shunt decreases organ perfusion, which correlates with such clinical problems as necrotizing enterocolitis and lower glomerular filtration rates. Clinical findings of a left-to-right shunt include collapsing pulses (widened pulse pressure), cardiomegaly, and a continuous murmur. Practically, this produces a continuing dependence on mechanical ventilation and an increasingly or persistently high oxygen requirement. Traditional therapy includes restricting IV fluids, administering digitalis, diuretics, and dopamine, and adding positive end-expiratory pressure (PEEP). All of these may influence the degree of shunting but do not directly promote closure.

Diagnosis is confirmed by 2D echocardiography, which is invaluable in assessing both patency and pressures. The hemodynamic significance of a PDA, rather than its patency, determines the course of treatment. The decision about when to close a PDA by medical or surgical means remains a clinical one.

NON-SURGICAL CLOSURE

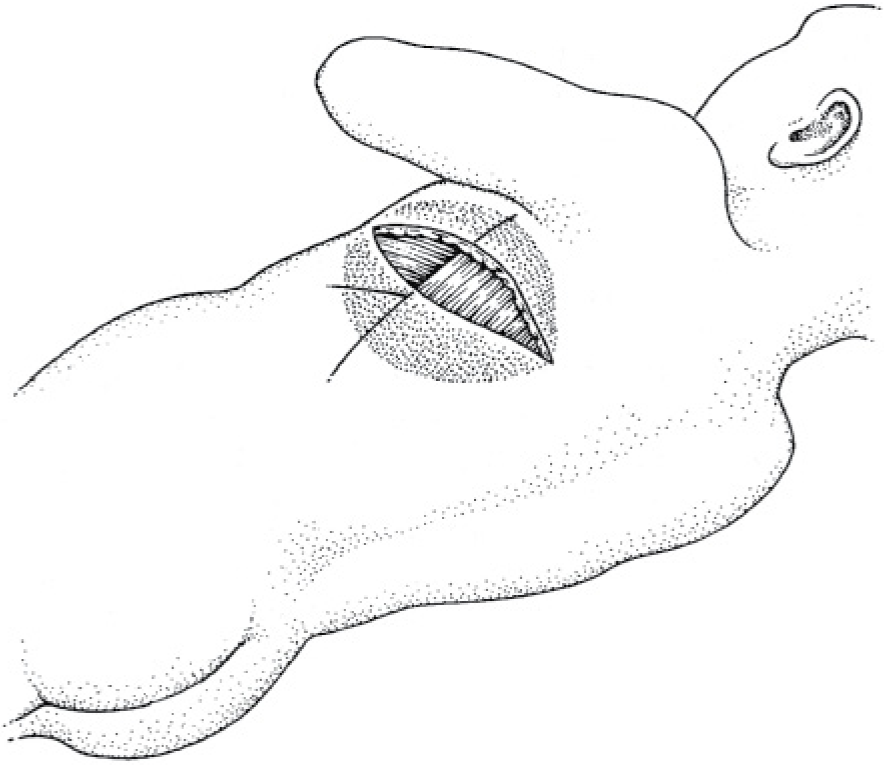
The treatment of choice for non-surgical closure of a PDA in most neonatal intensive care units has historically been indomethacin. Two or three courses are usually required to promote closure, and further administration beyond this rarely improves the success rate. Indomethacin tends to be less effective as the infant grows older. Contraindications to giving indomethacin include sepsis, necrotizing enterocolitis, azotemia, intraventricular hemorrhage, and coagulopathy. Newer data suggest that acetaminophen may be as effective as cyclo-oxygenase inhibitors in closing the PDA, with a lower reported incidence of complications. Acetaminophen does not seem to be effective after a failed course of ibuprofen. When contraindications to medical therapy exist, or if medical therapy has failed, surgical intervention is necessary for symptomatic PDA.

Endovascular occlusion is a developing alternative to surgical ligation, with percutaneous closure of a PDA now generally preferred over surgical ligation for infants over 4–6 kg. The introduction of smaller coils has recently permitted endovascular occlusion in much smaller infants – reported in extremely low birth weight (ELBW) babies below 1 kg) – but most series are limited to 1–2 kg. This procedure has been reported using a 4 Fr sheath placed via either a femoral venous or femoral arterial approach, with a 10% rate of arterial thrombus formation requiring anticoagulation reported with the arterial access approach. Most reported cases have been performed with general anesthesia. Endovascular occlusion in extreme prematurity is still considered a technically challenging procedure and not a routine alternative to surgical ligation due to associated technical complications related to vascular access and coil deployment.

OPERATION

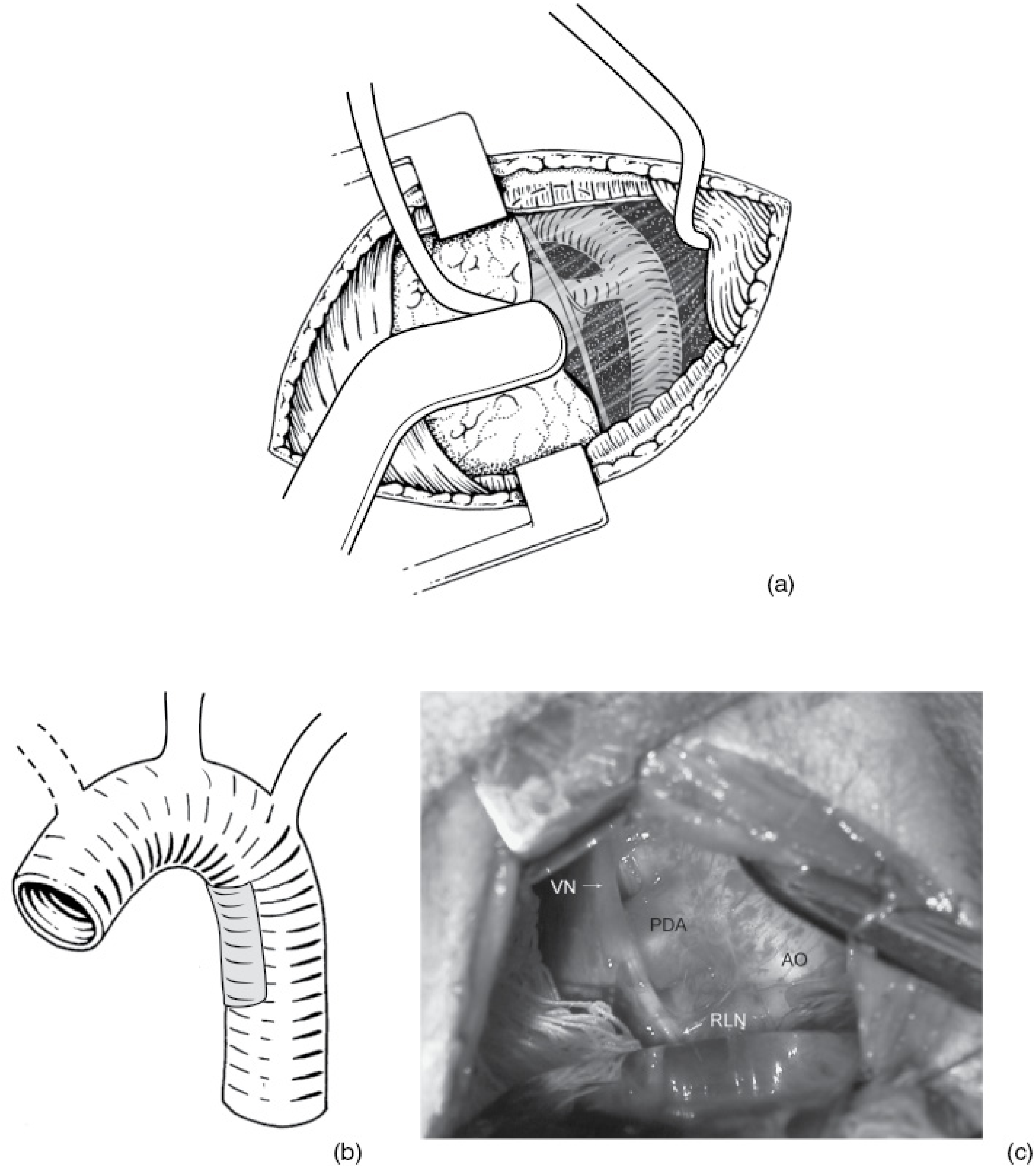
Open thoracotomy

The infant is placed in the lateral decubitus position with the right side down. The surgeon stands on the right side, facing the infant's abdomen. A headlight provides valuable illumination of the small operative field. A lateral incision is made and the subcutaneous layer is mobilized from the underlying muscles (**Figure 21.1**). The flimsy attachment of the anterior margin of the latissimus dorsi, which overlies the inferior edge of the serratus anterior, is incised. The latissimus is elevated and retracted posteriorly, and the serratus is freed and retracted anteriorly. It is unnecessary to divide any muscle to achieve exposure.



21.1

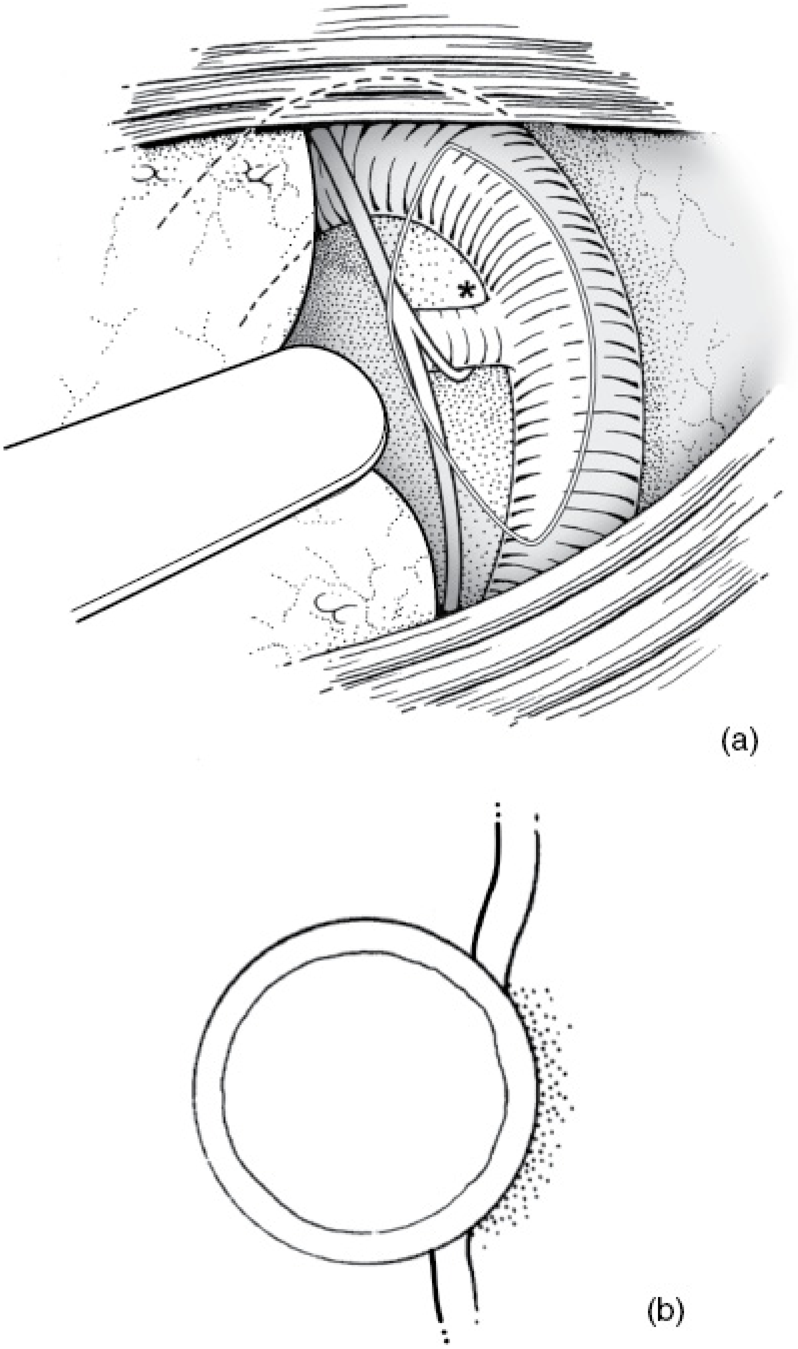
The third or fourth intercostal space is entered and a small self-retaining retractor (Finochetto) is placed. A second retractor (Gelpi) retracts the muscles in an anterior–posterior direction (**Figure 21.2a**). The ductus arteriosus may be as large as, or larger than, the aorta. Its exact position is variable; as it arises from the ventral side of the aorta, it may be located just inferior to or more distal from the aortic arch (**Figure 21.2b**). When mediastinal edema exists, and in the tiniest babies, the usual landmarks can be obscure. Identification of the vagus nerve and its recurrent branch always leads to the ductus arteriosus (**Figure 21.2c**: VN, vagus nerve; PDA, patent ductus arteriosus; AO, aorta; RLN, recurrent laryngeal nerve).



21.2a–c

The mediastinal pleura overlying the aorta is opened; a large superior intercostal vein often overlies the ductus and, if present, must be divided. A malleable retractor holds the lung anteriorly and inferiorly for exposure, but in unstable patients it is often necessary to allow intermittent lung expansion during surgery. The vagus nerve and recurrent branch are visible and are reflected anteriorly with the pleura.

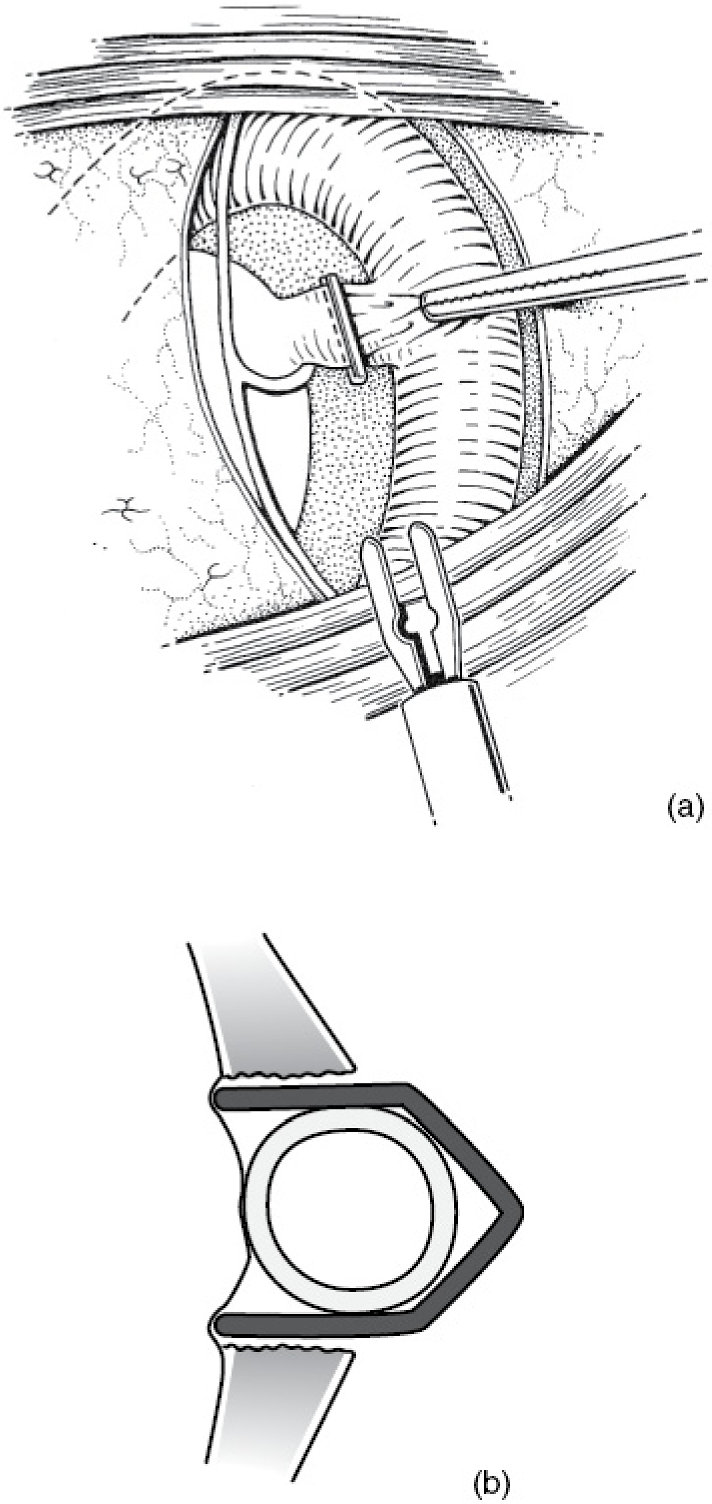
Adequate exposure of the ductus is achieved by incising and spreading the tissue just above and below the ductus. This delicate areolar tissue can be grasped with forceps, but care should be taken to avoid direct handling of the ductus, which is extremely friable. The use of tenotomy scissors enhances the accuracy of this dissection. The superior angle between the aorta and the ductus arteriosus (* in **Figure 21.3a**) is particularly vulnerable, and dissection here must be performed with great care. Another option is to open the pleura longitudinally over the center of the aorta at the level of the ductus. A flap can then be mobilized medially to expose the ductus and help protect the vagus and recurrent laryngeal nerves.



21.3a,b

It is not necessary to completely dissect and encircle the ductus unless it is too large for a metallic clip and requires ligation. If circumferential dissection is not performed, clear visualization and avoidance of the recurrent laryngeal nerve is essential (**Figure 21.3b**).

The adventitia of the aorta can be grasped and pulled away from the surgeon when necessary to provide enough length to place the clip. The tip of the metallic clip must extend beyond the margin of the ductal wall to ensure complete occlusion. The appropriate-sized metallic clip is placed closer to the aortic end of the ductus arteriosus (but not flush against the aorta) (**Figure 21.4a and b**). When ligation is necessary, a heavy ligature is safest, as it decreases the likelihood of cutting through the wall. It should be passed around the ductus arteriosus using a right-angled clamp.



21.4a,b

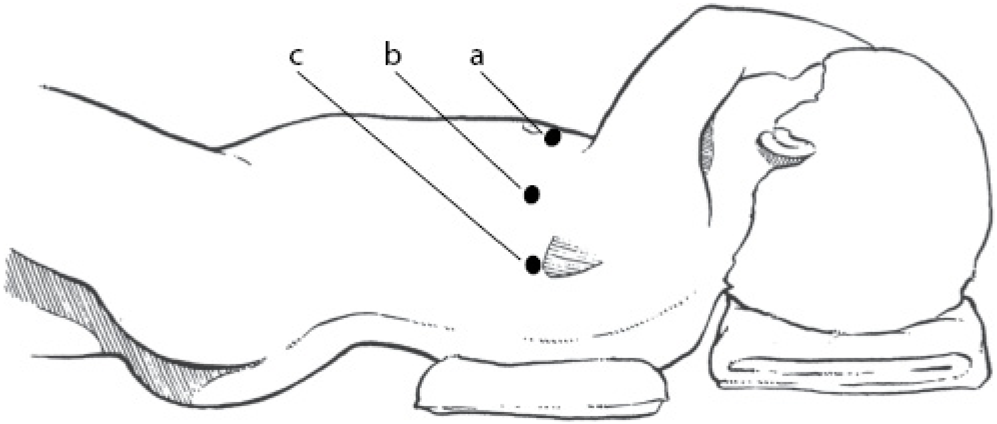
An alternative is to ligate the duct, usually with a 3/0 or 2/0 non-absorbable (silk) suture.

Several absorbable pericostal sutures are placed to appose the ribs and close the intercostal space gently. Since a muscle-sparing incision is used, no muscular sutures are needed. The lung is gently re-expanded with the ventilator and, although rarely needed, a small rubber catheter or thoracostomy may be placed into the pleural cavity and gentle suction applied. The catheter may be removed once the pericostal sutures have been tied, unless there is an obvious air leak. The intercostal muscle is not sutured, and a very fine skin closure is performed.

Thoracoscopic ductal ligation

Thoracoscopic PDA ligation is an alternative to open thoracotomy. It offers the theoretical advantages of better chest wall compliance, decreased risk of scoliosis, and cosmetic improvement.

For thoracoscopic PDA ligation, patients are placed in a modified right lateral decubitus position with the left side elevated approximately 30 degrees. Three ports are used (**Figure 21.5**) with a 4 mm port placed just posterior to the tip of the scapula for the camera. A 3 mm port is placed anteriorly approximately one or two interspaces above and a 3 mm port, which is changed to a 5 mm port for clip application, is placed posteriorly one or two interspaces below the camera port. All patients are monitored with pre- and post-ductal pulse oximetry. The lung is retracted medially and the parietal pleura overlying the proximal descending aorta and PDA are dissected free. There is usually a small vessel over the PDA which should be sealed initially to prevent bleeding with the dissection. The recurrent laryngeal nerve can clearly be visualized and care must be taken throughout to avoid injury. The PDA is then carefully dissected free on both sides. The ductus is test-clamped with an atraumatic grasper to ensure no negative hemodynamic events occur with compression of the structure. The posterior port is changed to a 5 mm port and the clip applier is introduced. A clip can then be applied.



21.5

Lung compliance and reduced visualization due to bleeding are the main reasons for conversion to an open approach.

POSTOPERATIVE CARE

In many centers, the operation is performed in the neonatal intensive care unit, as transportation of these tiny, unstable infants can be challenging. The operation should be completed in less than 30 minutes. After surgery, lung re-expansion is confirmed by chest radiography. The infant should recover quickly from the short-acting IV agents. The hemodynamic response of interrupting a large left-to-right shunt is immediate, but the gratifying increase in blood pressure may require prompt treatment to avoid central nervous system hemorrhage.

COMPLICATIONS

Postoperative complications are uncommon, but chylothorax, transient vocal cord paralysis, and incomplete occlusion have been reported, in addition to the usual sequelae of any thoracic surgical operation. If the ductus is torn, or there is inadvertent ligation of the left pulmonary artery or aorta, the baby is unlikely to survive.

OUTCOME

Improvement in cardiopulmonary function starts immediately, depending on the volume of flow through the left-to-right shunt and the preoperative status of the heart and lungs. Many studies have tried to compare the results of indomethacin versus surgical ligation but these fail to be convincing because the patient groups are different, and many babies undergo surgery only because medical therapy has failed. Because the method of ductal closure appears to have little influence on overall survival, accurate comparison of these modalities is difficult.

VIDEO-ASSISTED THORACOSCOPIC APPROACH

Video-assisted thoracoscopic PDA ligation has been shown to be feasible. Large series of patients undergoing this technique report similar outcomes with the standard advantages of thoracoscopy over thoracotomy being noted. These include improved cosmesis, reduced pain, shorter recovery time, and better visualization. While there have been reports of successful treatment of premature neonates, this procedure is generally reserved for the pediatric population and is often compared to percutaneous techniques, such as coil occlusion. The application of thoracoscopic ligation in small premature neonates is limited due to clinical stability of the patient, operative mechanics such as instrument size, and visualization on high-frequency ventilation, as well as the risk and impact of injury to the ductus, aorta, and pulmonary artery.

VIDEO

OPS_8e_video_21.1 Thoracoscopic PDA ligation

FURTHER READING

El-Mashad AE, El-Mahdy H, El Amrousy D, et al. Comparative study of the efficacy and safety of paracetamol, ibuprofen, and indomethacin in closure of patent ductus arteriosus in preterm neonates. *Eur J Pediatr* 2017; 176: 233–40.

Hines MH, Raines KH, Payne RM, et al. Video-assisted ductal ligation in premature infants. *Ann Thorac Surg* 2003; 76: 1417–20.

Rodríguez Ogando A, Planelles Asensio I, de la Blanca ARS, et al. Surgical ligation versus percutaneous closure of patent ductus arteriosus in very low-weight preterm infants: Which are the real benefits of the percutaneous approach? *Pediatr Cardiol* 2018; 39(2): 398–410.

22

Thoracoscopic sympathectomy

ASHER PRESSMAN

BACKGROUND

Primary hyperhidrosis (PH) is a relatively common chronic condition that causes excessive sweating in various parts of the body. It is caused by hyperreactivity of the sympathetic system resulting in excessive stimulation of the eccrine sweat glands. The most commonly affected areas are the palms, axillae (axillary hyperhidrosis, AH), and soles of the feet; however, additional areas can also be affected, such as the face and scalp. Primary palmar hyperhidrosis (PPH) is the most common variant of PH but is of unknown etiology.

The prevalence of the condition ranges from 1–2% in the West to 4–5% of the population living in the Middle and Far East. It occurs in all seasons regardless of the body heat control needs. Symptoms are exacerbated during hot and humid weather, stress situations, anxiety, and nervousness.

There is no gender difference and a family history of primary hyperhidrosis is very common.

Clinical manifestations of PPH include wet and cold palms accompanied by change of color from pale to blush. They usually appear before the third decade of life, often during childhood and adolescence. Severe palmar perspiration can significantly impair patients' quality of life. It has serious social and emotional effects; in many cases patients are no longer able to perform routine everyday activities, do schoolwork, or engage in interpersonal social relations.

As PPH is not a self-limiting condition, treatment is recommended even in children and adolescents in order to reduce suffering.

Various medical treatment options are available and include topical agents, systemic anticholinergic medications, and local therapy. Examples include topical treatment with astringent solutions, aluminum chloride preparations, and 2% glycopyrrolate solution; oral medications such as glycopyrrolate, oxybutynin, and methanthelinium bromide. The use of tap water iontophoresis, acupuncture, and intradermal injection of Botulinum toxin (e.g. Botox™ which lasts 3–6 months) may be useful in some patients. Psychotherapy and hypnosis may also aid in relief of the symptoms.

Endoscopic thoracic sympathectomy (TS) was first described in 1942 by Squadron Leader John Hughes from Sheffield, UK. With the emergence of video-assisted technology it has become the standard surgical approach and is the only permanent cure for both PPH and AH.

OPERATION

Position of patient

Standard single-lumen or double-lumen endotracheal intubation is used according to preference.

The patient lies supine on the operating table with the thorax elevated by about 25 degrees or in a 25-degree anti Trendelenburg position and both arms are abducted to 90 degrees. The aim is to deal with both sides in one operation.

We use a one-port technique, for both the camera and the diathermy probe. A 5 mm or 10 mm laparoscope is used according to the age and size of the patient, with a working channel at a zero-degree view.

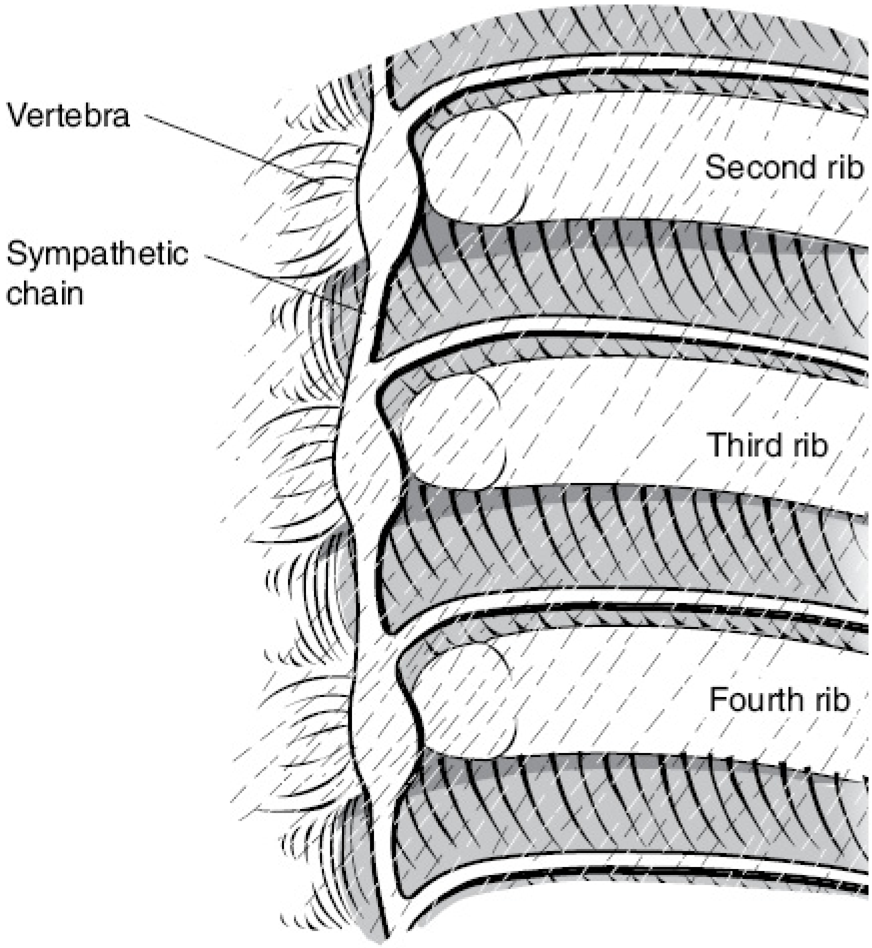
Incision

Local anesthesia is infiltrated and an incision made in the anterior axillary line at the third or fourth intercostal interspace. Blunt dissection using an artery forceps allows penetration of the pleura. A 5 mm or 10 mm trocar is then inserted and gas administered to induce pneumothorax to about 10 mmHg. During the very short time of pleural penetration and trocar insertion the patient is not ventilated to prevent lung injury.

Procedure

The aim of the procedure is to cut the sympathetic chains at the level of the second, third, and fourth ribs (for AH).

Once lung collapse has been achieved, a 3 mm or 5 mm probe or hook is introduced through the working channel and under vision. The sympathetic trunk can be identified by sight and by physically sensing a “string-like” feeling with the probe. The sympathetic chain runs vertically, parallel to the vertebral column, on both sides, overlying the neck of the ribs in the upper tubercle region, under the parietal pleura (**Figure 22.1**).

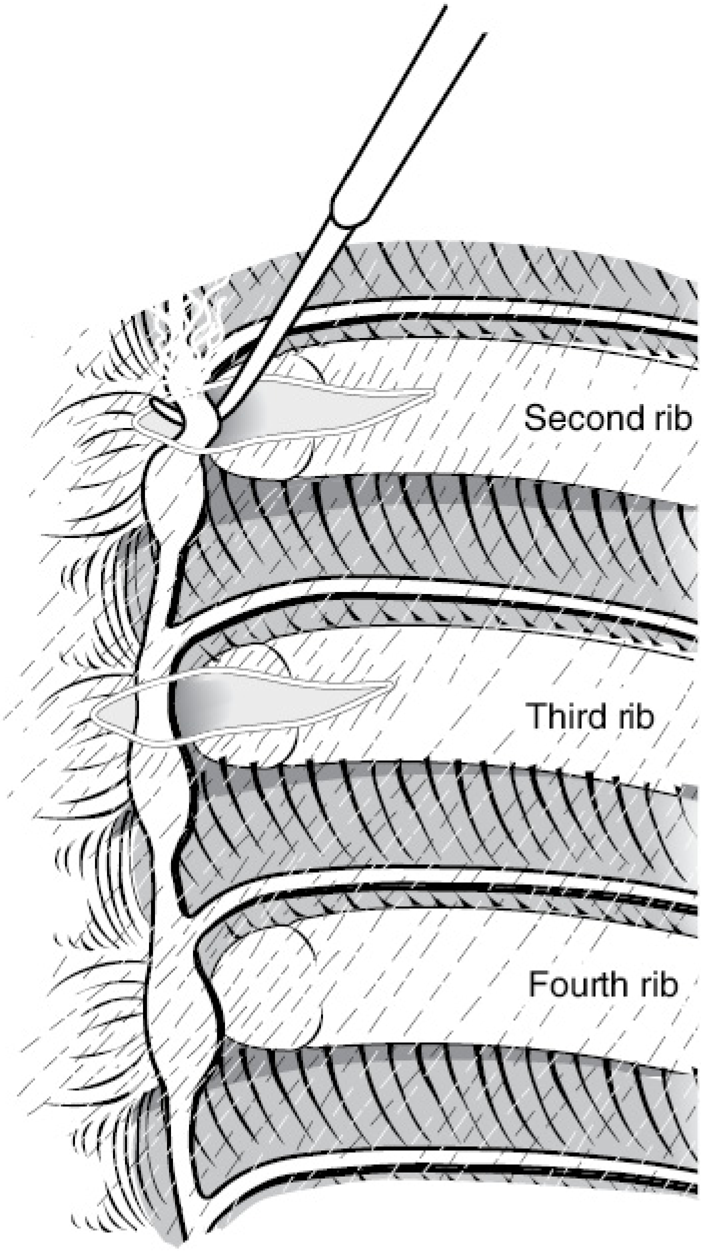


22.1

The first rib and stellate ganglion are covered by Sibson's fascia and not visible from within the chest cavity. The most proximal rib seen is the second, with the second ganglion on it. The dissection starts here, moving down toward the third or fourth rib.

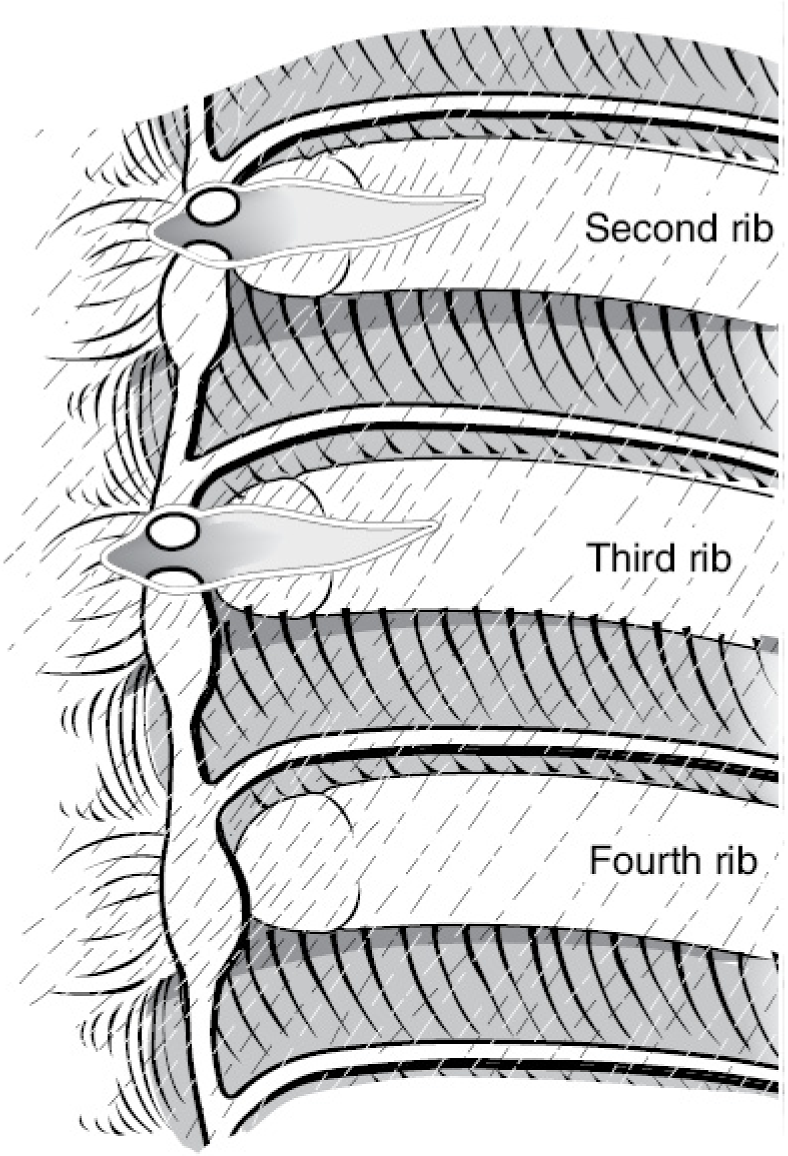
A 3 mm or 5 mm diathermy hook is inserted through the working channel under vision. Hook diathermy dissection of the pleura on both sides, parallel to the sympathetic ganglia, on levels T2 and T3 and, in some cases of AH, also T4, creates a window from which the second, third, and fourth ganglia can be seen, elevated, and dissected. This is most easily done by pressing the diathermy hook down onto the bone of the ribs neck, parallel to the chain.

Through the pleural windows created, the sympathetic chain is dissected by the hook, elevated from the bone, and then cut using the diathermy (Figure 22.2).


Second rib
Third rib
Fourth rib

22.2

Once the sympathectomy is complete (**Figure 22.3**), the lung is reinflated under vision and the procedure is repeated on the contralateral side. Chest drains are not routinely used. Skin closure is achieved with tissue glue.



22.3

Side effects

Horner's syndrome may occur because of transmitted energy damaging the stellate ganglion and has been reported in <2% of cases.

Compensatory sweating (CS) particularly of the trunk is very common in both adults (>85%) and children (60%) but seems to be tolerated better by children than adults. Occasionally, this symptom becomes disabling in adults. CS develops immediately post-operation and can worsen during the first months after the procedure. However, it often improves significantly within 2 years post-surgery. Some studies suggest that preserving the T2 ganglion may reduce the risk of this particular complication.

VIDEO

OPS_8e_video_22.1 Thoracoscopic sympathectomy

FURTHER READING

Cerfolio RJ, De Campos JR, Bryant AS, et al. The Society of Thoracic Surgeons expert consensus for the surgical treatment of hyperhidrosis. *Ann Thorac Surg* 2011; 91: 1642–8.

Lin T-S. Transthoracic endoscopic sympathectomy for palmar and axillary hyperhidrosis in children and adolescents. *Pediatr Surg Int* 1999; 15: 475–8.

Ojimba TA, Cameron AEP. Drawbacks of endoscopic thoracic sympathectomy. *Br J Surg* 2004; 91: 264–9.

Romero FR, Haddad GR, Miot HA, Cateneo D.C Palmar hyperhidrosis: Clinical, pathophysiological, diagnostic and therapeutic aspects. *An Bras Dermatol* 2016; 91:716–25.

Steiner Z, Cohen Z, Kleiner O, et al. Do children tolerate thoracoscopic sympathectomy better than adults? *Pediatr Surg Int* 2008; 24: 343–7.

23

Thymectomy

STEVEN S. ROTHENBERG

HISTORY

The first thymectomy was performed by Sauerbrunchin in 1911 in a woman with hyperthyroidism and myasthenia gravis (MG). The thymectomy was performed to treat her hyperthyroidism, but both conditions improved temporarily. Further understanding of the surgical approach to the thymus and its relationship to systemic disease was documented by Blalock and his colleagues when they successfully removed the thymus in a young woman with a thymic cyst and MG. She eventually underwent complete remission. They later reported the removal of an anatomically normal thymus with clinical improvement of the disease and this became the basis for the surgical management of myasthenia.

Thymectomy is an established therapy in the management of generalized MG and can be implemented as a supplement to medical therapy, or in cases where medical therapy has failed to improve symptoms. The course of the disease is highly variable and the severity of the disease often changes. The timing of thymectomy is controversial, but most agree that pediatric patients who present with the disease do better with thymectomy.

The other indication for thymectomy is thymic tumors, and these are often associated with immunodeficiency syndromes as well. Nearly 50% of patients with thymic tumors present with symptoms of MG. The majority of others are asymptomatic but may present with chest pain, dyspnea, cough, fatigue, and weight loss. These tumors can include benign thymic cysts (10%) and thymomas.

PRINCIPLES AND JUSTIFICATION

It is well known that the thymus is necessary for the development of cellular immunity and T lymphocytes. However, once past infancy the thymus seems to have little function and removal causes no alterations in immune function of normal individuals. The role of the thymus in MG is now clearly recognized and thymectomy is an established and accepted therapy as part of the overall treatment plan. Myasthenia is an autoimmune disorder of the postsynaptic nicotinic acetylcholine receptor and manifests as weakness and fatigue of skeletal muscles. Ptosis, diplopia, dystonia, and loss of facial expression are often common early findings. More severe symptoms include severe weakness and even respiratory compromise. Initial medical therapy consists of drugs aimed at blocking the effect of antibodies at the neuromuscular junction. First-line drugs are anticholinesterases, such as pyridostigmine and mestinon. Steroids and other immunosuppressants are used in more severe cases. Plasmapheresis has also been used to filter out antibodies to the acetylcholine receptors.

INVESTIGATIONS

The workup for patients with a thymic mass is relatively straightforward. In the case of large masses, the patient may present with symptoms of respiratory compromise or distress from tracheal compression. A routine chest X-ray may show evidence of mediastinal widening or shift.

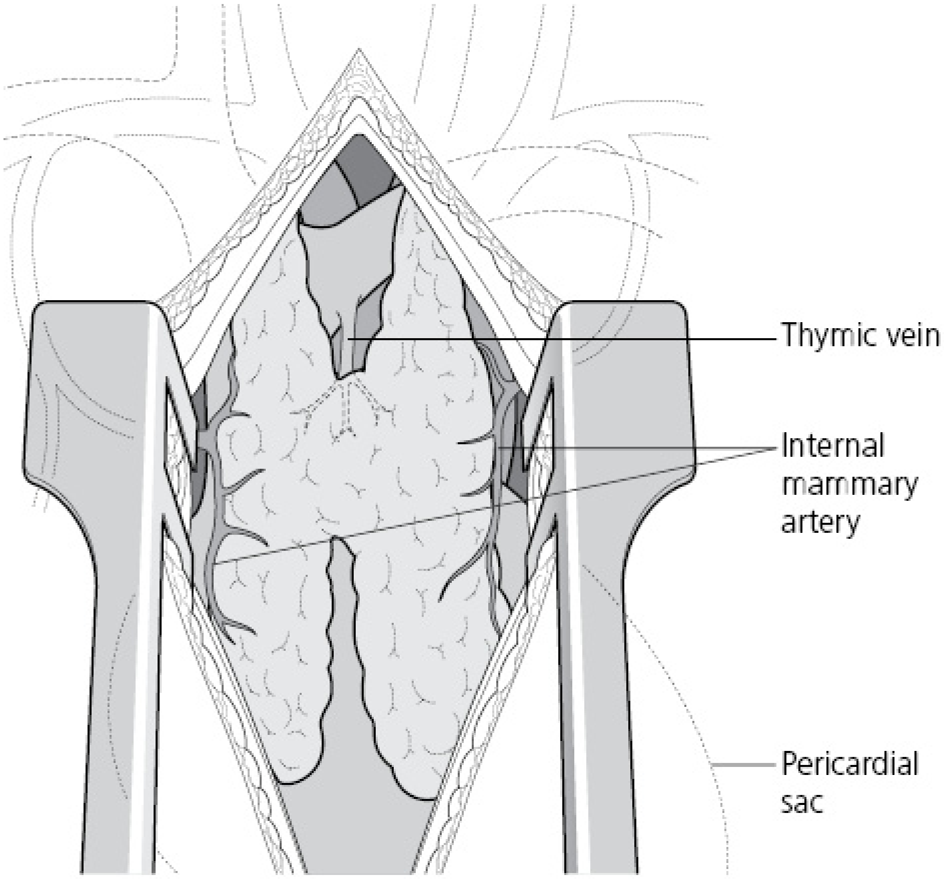
A computed tomography (CT) scan or, in some cases, a magnetic resonance imaging (MRI) scan are usually obtained to determine the extent of the mass and to plan the best surgical approach. No specific investigations are required preoperatively in cases of MG; the preoperative and postoperative management, however, are more critical.

Most patients with MG are on anticholinesterase therapy and may be on steroids as well. These medications need to be maximized and monitored closely both pre- and postoperatively. Depending on the severity of muscle weakness and respiratory compromise, plasmapheresis may be indicated preoperatively to try to diminish postoperative respiratory issues. Intravenous immunoglobulin therapy can also be instituted to diminish the necessity for postoperative ventilation.

OPERATIONS

There are a number of different surgical approaches to thymectomy and the decision of which one to use depends on the disease process and surgeon preference. However, with perhaps the exception of a giant anterior mediastinal mass, a thoracoscopic approach is generally the most preferable in terms of overall surgical exposure of the gland and the significantly decreased morbidity for the patient. Other approaches include a full median sternotomy, a partial sternal split, or a transverse cervical incision. The anatomy of the thymus often plays a role in surgeon bias when contemplating the surgical approach, especially in cases of MG where a complete resection of all thymic tissue is required.

The thymus lays in the anterior mediastinum and overlies the pericardium and the great vessels. The thymus is a bilobed structure with upper and lower poles resembling an H (**Figure 23.1**). The upper poles extend up into the neck and are attached to the thyroid gland by the thyrothymic ligament. The lower poles extend down over the pericardium. The hilum of the thymus lies over the left innominate vein. The venous drainage of the thymus is usually through one or two small branches to this vein. The arterial supply comes in laterally off the internal mammary or thyroid arteries.



23.1

The extensive length of the thymus both above and below the thoracic inlet is what caused many surgeons to advocate a median sternotomy, as there is concern that a more limited incision made complete resection difficult if not impossible. A thoracoscopic approach eliminates this concern as visualization of the entire anterior mediastinum, including up into the neck, is facilitated by use of the thoracoscope.

Anesthesia

The keys to anesthesia are really the preoperative preparation as already mentioned. The greatest concern is being able to extubate the patient postoperatively because of the weakness of the respiratory muscles.

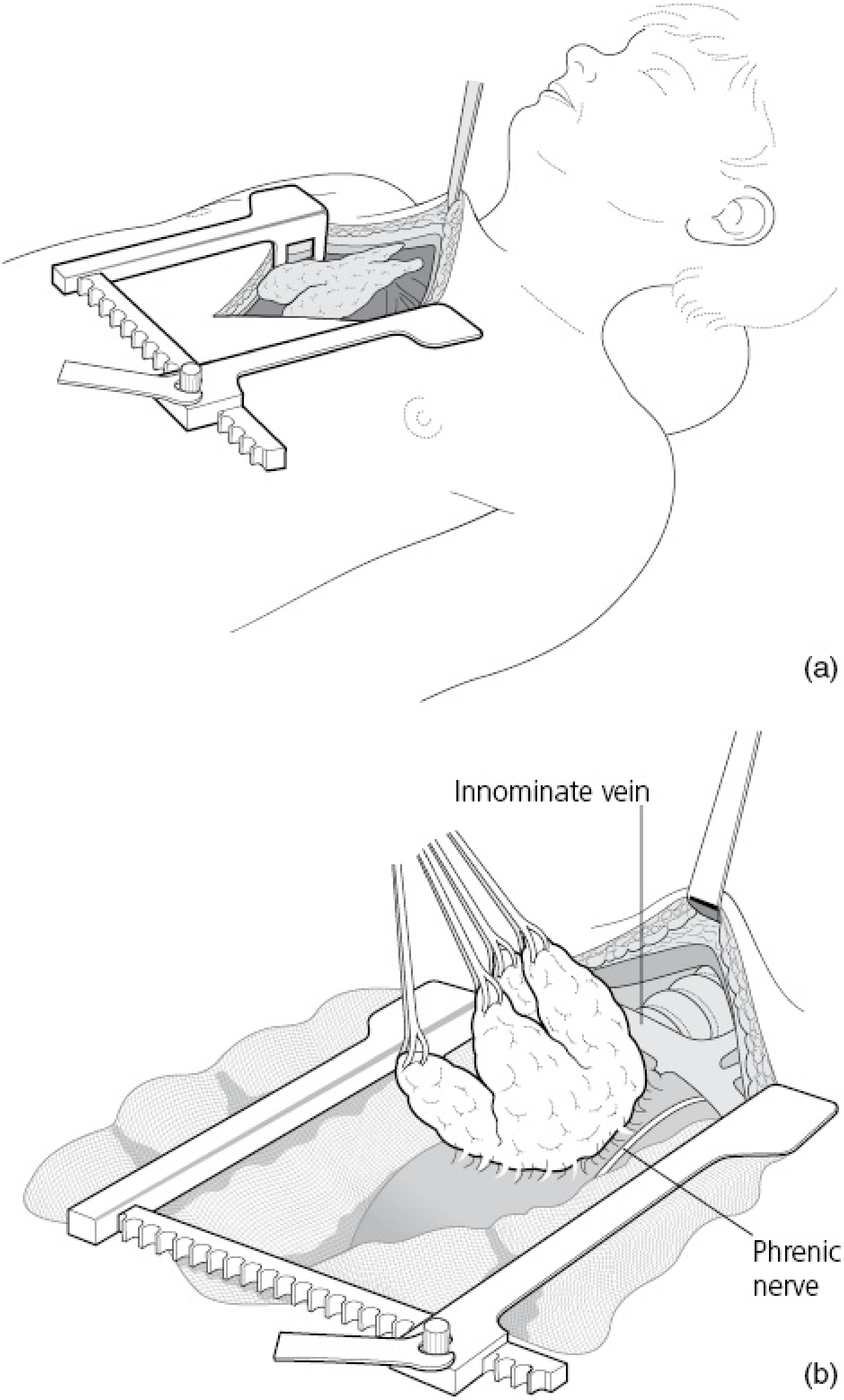
If a median sternotomy is used, this is a significant concern. A cervical or a thoracoscopic approach is associated with significantly less pain and these patients are usually more easily extubated.

Some advocate single-lung ventilation during a thoracoscopic thymectomy and this does ease access and improve exposure. The author prefers to approach the thymus through the left chest so generally uses a mainstem intubation of the right bronchus to achieve this. Other options include the use of a double-lumen endotracheal tube or bronchial blocker.

Adequate exposure can also be obtained by insufflation with CO_2 alone. Generally, a pressure of 4–6 mmHg is adequate to collapse the lung sufficiently to allow safe dissection of the thymus.

Sternotomy

A sternotomy is a relatively standard and straightforward approach. The patient is in the supine position with the arms at the side. The incision runs in the midline from the sternal notch to the xiphoid. The sternum is split using an oscillating sternal saw and exposure of the thymus is obtained by use of a sternal retractor (**Figure 23.2a**). The dissection is relatively easy and can begin with either the lower or upper horns. The thymus is relatively easily dissected off the pericardium and great vessels. Care must be taken to protect the phrenic nerve, which runs along the lateral border of the thymus, down the pericardium. Care should also be taken to avoid entering the pleural cavity if possible.


(a)
Innominate vein
Phrenic nerve
(b)

23.2a,b

The vascular supply is easily addressed as the arterial branches come in anteriorly and laterally off the internal mammary, and the veins are easily exposed by rolling the thymus medially off the left brachiocephalic vein once the upper and lower pole are mobilized (**Figure 23.2b**). Complete excision of the thymus should be guaranteed with this approach as the entire gland is exposed.

Transverse cervical incision

This approach has been advocated to avoid the morbidity of a sternotomy, especially the respiratory compromise. With the advent of thoracoscopy, there is probably little advantage to this technique, although it should avoid any disruption of the pleural space.

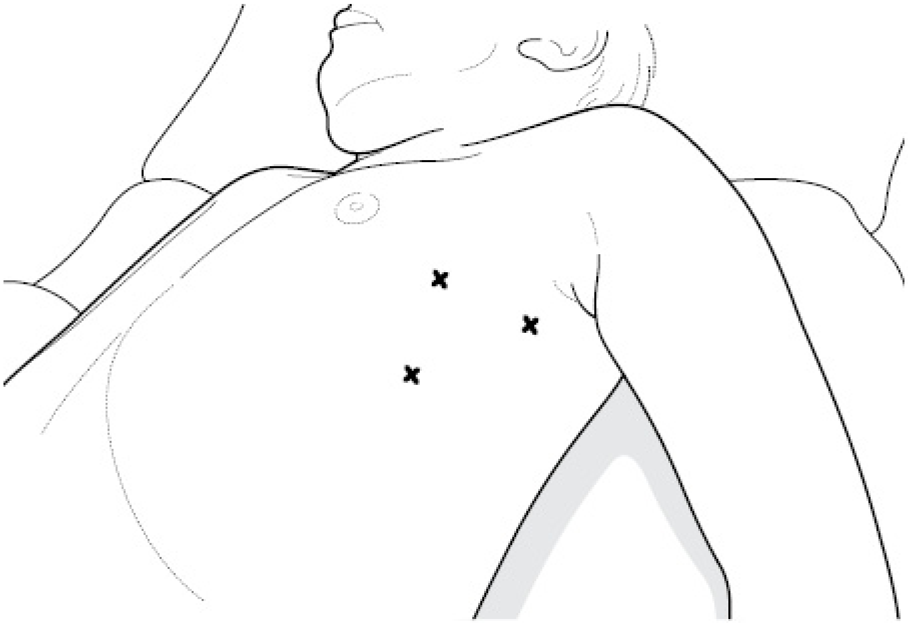
A transverse incision is made just above the sternal notch extending from the anterior border of the sternocleidomastoid muscle on one side to the other. Dissection is carried down in the midline to the trachea. The thymic isthmus and the upper horns of the right and left lobe should be easily identified. Dissection of the upper poles is under direct vision and straightforward. Once the upper poles are mobilized, gentle cranial traction is placed on the gland, and the body and lower poles are teased up into the incision. The thymic and brachiocephalic veins must be carefully looked for and the thymic vein ligated as it is easy to avulse the vein through this approach. If this happens, it is extremely difficult to regain vascular control through this limited incision. The key is constant gentle traction without tearing the gland.

The major criticism of this approach is that adequate exposure of the lower horns cannot be obtained and that there will be retained thymic tissue left in the lower anterior mediastinum. The phrenic nerves are also not visualized using this approach.

Thorascopic thymectomy

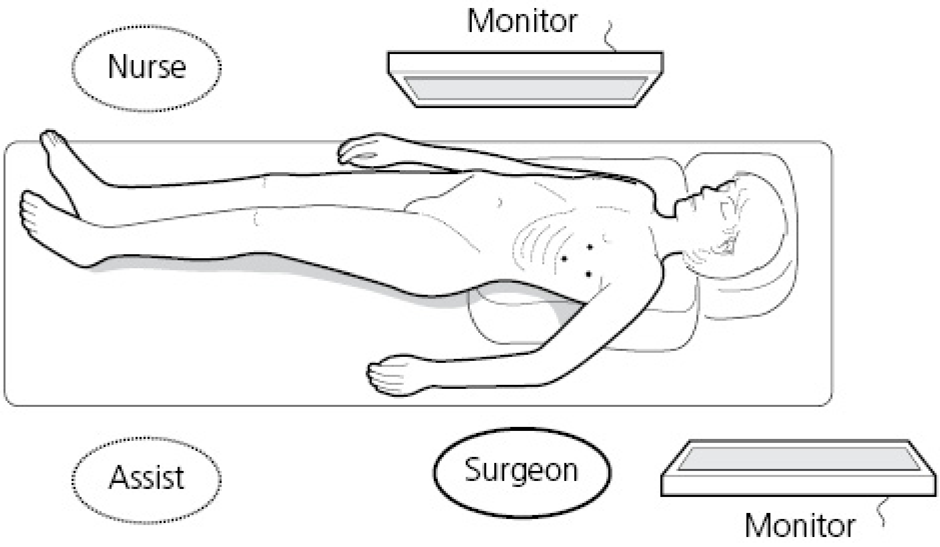
This is by far the most common approach today and avoids the pitfalls of the other techniques.

The patient is in a modified supine position with the left side elevated 20–30 degrees to allow access to the lateral chest for trocar insertion (**Figure 23.3**).



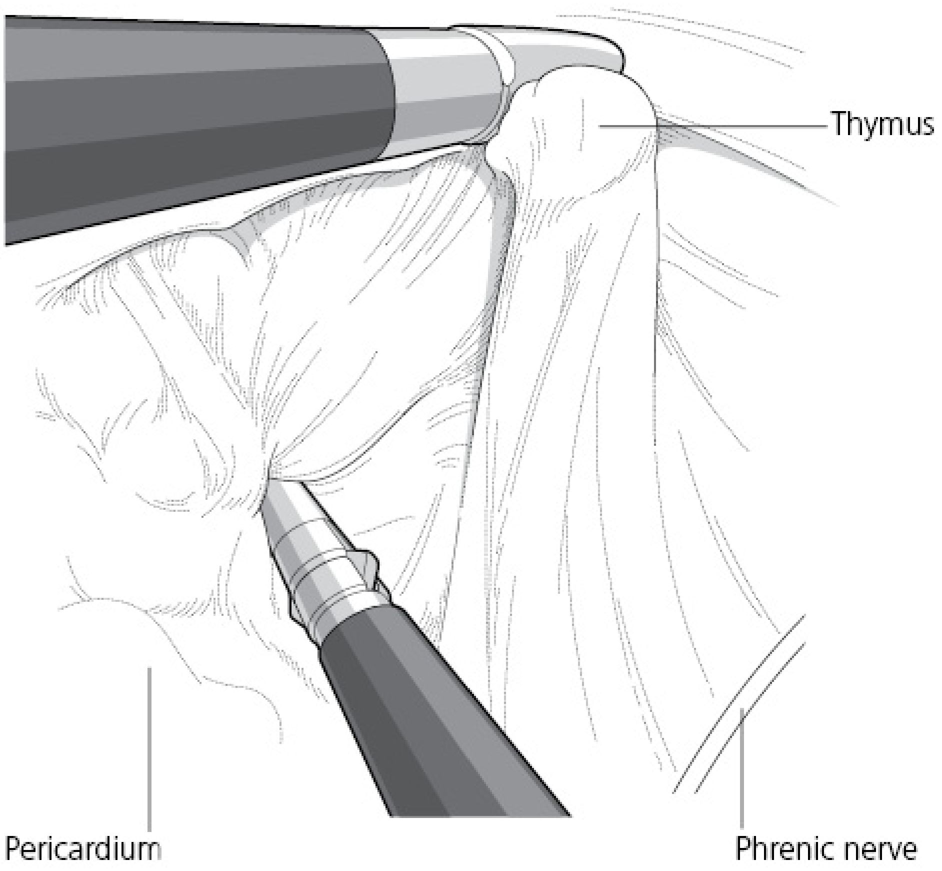
23.3

This position allows gravity to retract the lung posteriorly, exposing the anterior mediastinum. The arm is placed at the side, slightly hyperextended, or can be draped across the upper chest. The surgeon and assistant stand on the patient's left with the monitor directly across from them (**Figure 23.4**).



23.4

The chest is first insufflated with CO_2 using a Veress needle to ensure adequate collapse. This is done in the anterior axillary line in the fourth or fifth inner space. A 5 mm trocar is then inserted in this site and a 5 mm 30-degree scope is used for the procedure. Two other 5 mm ports are then placed superiorly and inferiorly to the camera port as shown in **Figure 23.5**.

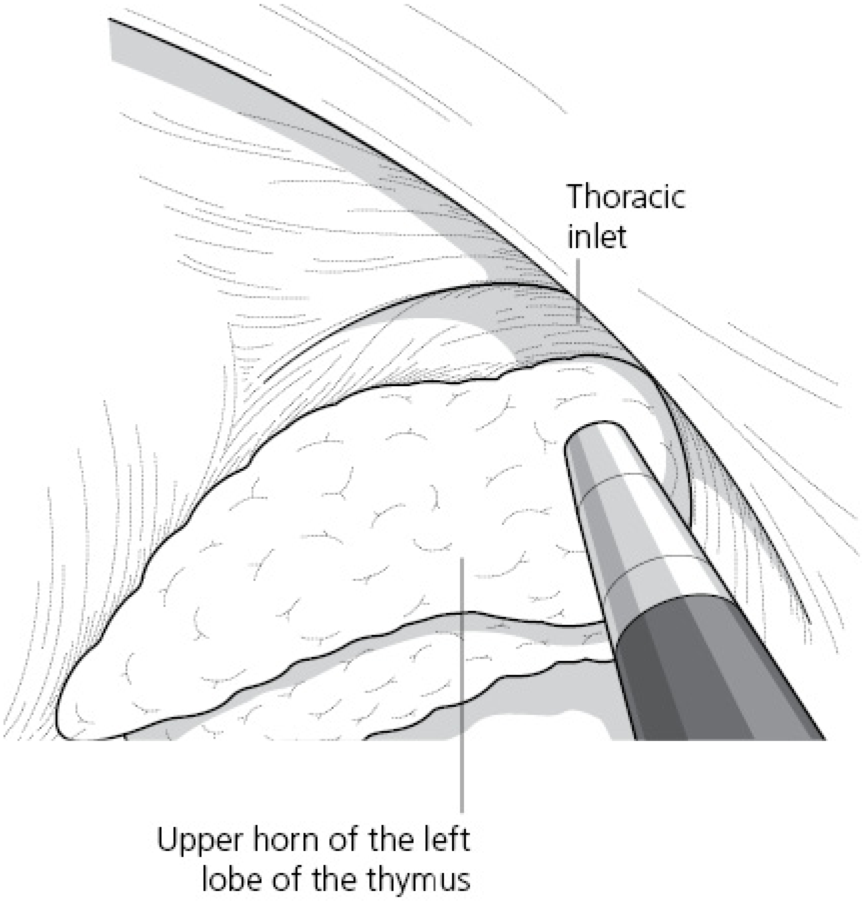


23.5

The thymus can easily be visualized lying on the pericardium.

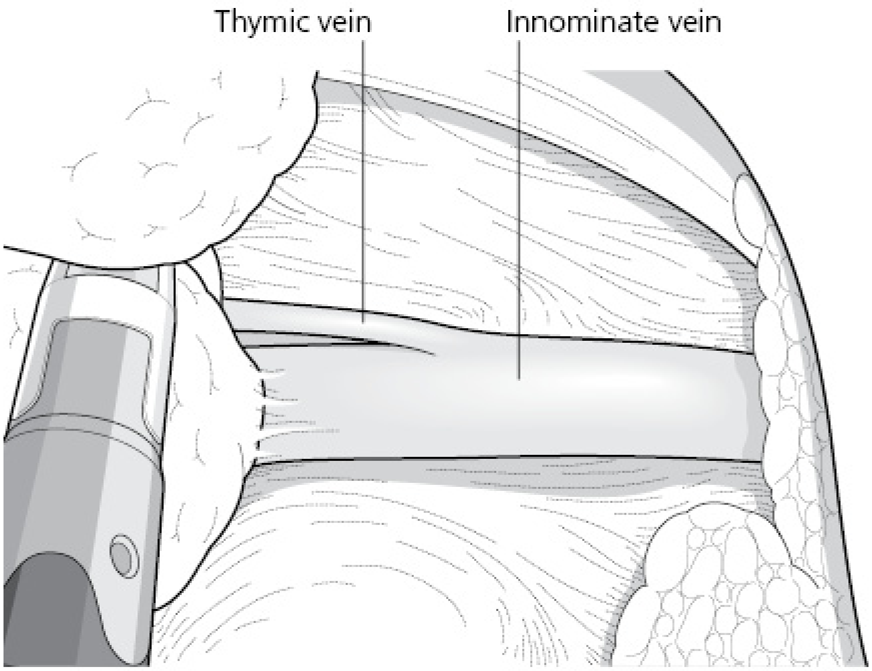
With a Maryland in the left hand and a scissor or tissue sealer/divider in the right, the pleura overlying the lateral border of the lower pole of the thymus is incised. Prior to this, the left phrenic nerve needs to be identified and care taken to avoid injury to this structure during the entire dissection.

Once the parietal pleura is open, the lower pole is freed by gently dissecting it off the pericardium and retracting it anteriorly and medially (**Figure 23.6**). Dissection is continued superiorly along the lateral border of the thymus up toward the upper pole. Small arterial branches may be encountered, but these are easily taken with the tissue sealer or cautery. Gentle traction is placed on the upper pole and blunt dissection is used to free it from the thyrothymic ligament. The thoracoscope provides for direct vision high into the neck through the thoracic inlet, allowing for complete dissection of the upper pole.


Thoracic inlet
Upper horn of the left lobe of the thymus

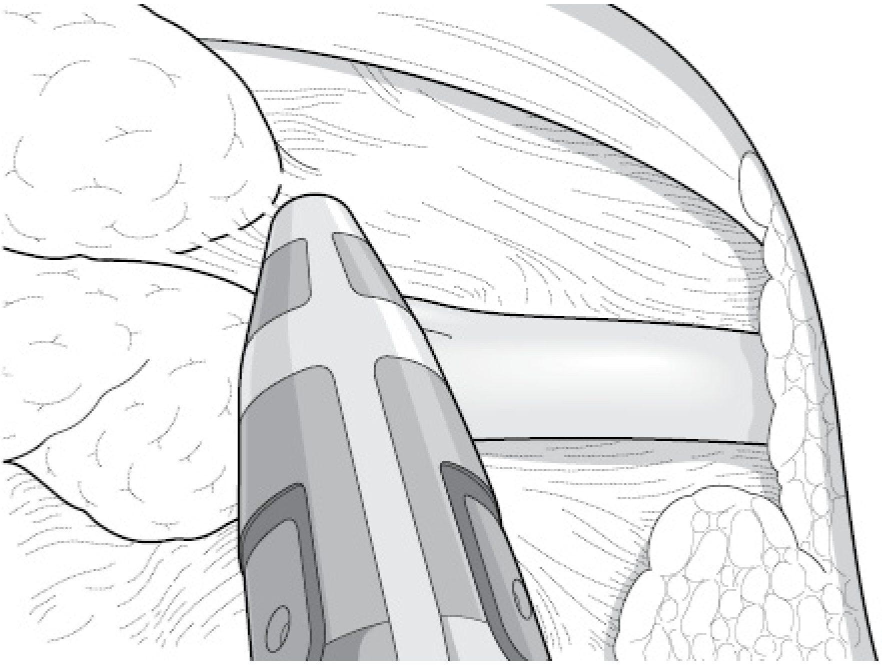
23.6

With the upper and lower poles mobilized, the isthmus is retracted medially exposing the back of the thymus gland and the left innominate vein. The thymic vein(s) is usually easily identified and it is further dissected out with gentle blunt dissection (**Figure 23.7**).


Thymic vein
Innominate vein

23.7

Once mobilized, the vein can be clipped and divided or sealed with the tissue sealer and divided (**Figure 23.8**).



23.8

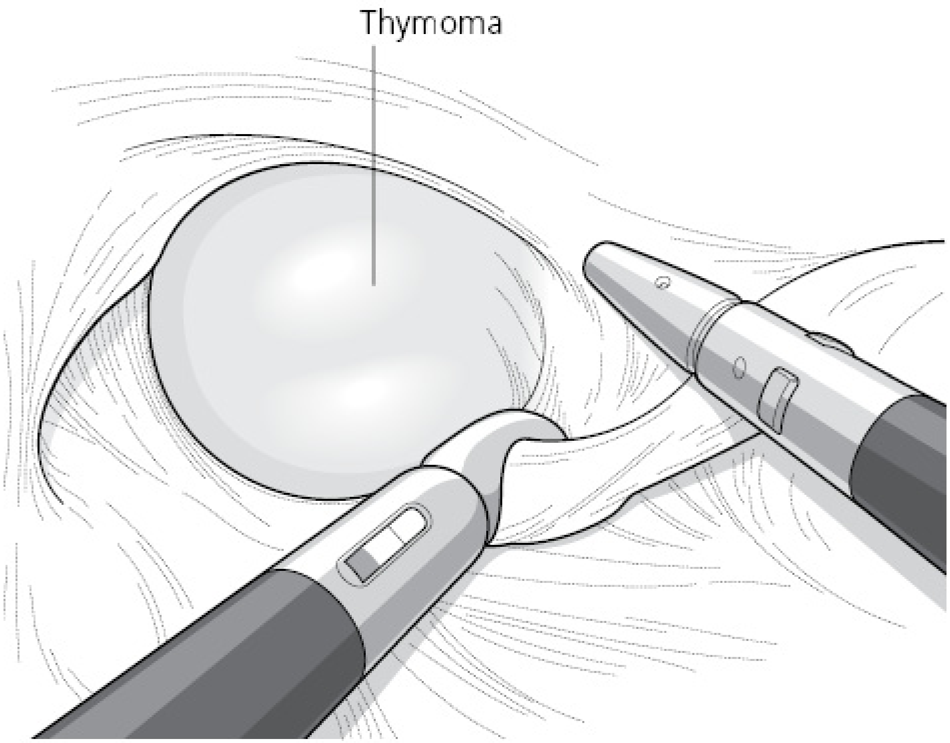
With this maneuver completed, the left lobe is now retracted into the left chest, pulling the right lobe of the thymus medially. The right upper and lower horns are gently dissected off their fascial attachments by blunt and sharp dissection. It should now be possible to dissect the thymus out of the right side of the mediastinum without entering the right pleural space.

The only danger to this part of the dissection is possible injury to the right phrenic nerve, as it cannot be easily visualized. The key to avoiding injury to this nerve is to maintain steady traction on the thymus, pulling it into the left chest, and using mostly blunt dissection for this part of the procedure. This pulls the gland away from the nerve and protects it.

Once the thymus is completely free, it is placed in an endoscopic specimen bag and brought out through a slightly enlarged trocar site.

The CO_2 insufflation is stopped and the right lung reinflated using the trocar to evacuate the pneumothorax. The last trocar is then withdrawn under positive pressure and an occlusive dressing is applied. No chest tube is necessary.

In cases of tumors, the same approach is used although the side of chest entry may vary depending on where the tumor lies (**Figure 23.9**).



23.9

The ability to resect the tumor thoracoscopically depends on the size of the mass and the surgeon's experience.

POSTOPERATIVE CARE

The patient should be extubated at the end of the procedure and simply requires aggressive chest physiotherapy. No chest tube is left in place and there will likely be a small residual pneumothorax on the chest X-ray, but this will resolve spontaneously over the next 24–48 hours.

Most patients are discharged on the first postoperative day. In cases of MG, the patient's medications are tapered over the next several weeks.

FURTHER READING

DeFilippi VJ, Richman DP, Ferguson MK. Transcervical thymectomy for myasthenia gravis. *Ann Thorac Surg* 1994; 57(1): 194–7.

Kogut KA, Bufo AJ, Rothenberg SS, Lobe TE. Thoracoscopic thymectomy for myasthenia gravis in children. *J Pediatr Surg* 2000; 35: 1576–7.

Seguier-Lipszyc E, Bonnard A, Evrard P, et al. Left thoracoscopic thymectomy for myasthenia gravis in children. *Surg Endosc* 2005; 19: 140–2.

PART IV

Abdominal

24

Inguinal hernia, hydrocele, and other hernias of the abdominal wall

SOPHIA ABDULHAI AND TODD A. PONSKY

INGUINAL HERNIA AND HYDROCELE

HISTORY

The first reference to hernia repair in children has been credited to Celsus, who in AD 25 recommended removal of the hernia sac and testes through a scrotal incision. The first accurate description of inguinal hernias was made by Sir Percivall Pott writing in "A Treatise on Ruptures" published in 1756. Czerny advocated high ligation of the hernial sac through the external ring in 1877, which was superseded at least in adults by opening the canal, narrowing the internal ring and repair of the posterior wall as advocated by Edoardo Bassini in the 1880s.

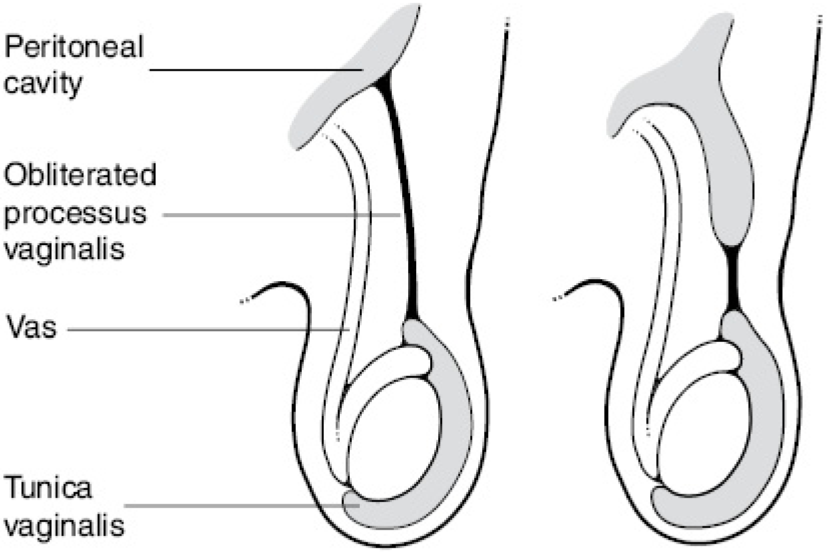
In 1912, Turner showed that simple high ligation of the sac at the level of the internal ring was the only procedure necessary in most children. The concept of day-case outpatient surgery began in the 1920s by Gertrude Herzfeld in Edinburgh, and early repair in infancy was recommended by William Ladd and Robert Gross in 1941 from the Boston Children's Hospital.

The first laparoscopic inguinal hernia repair was performed by Phillipe Montupet in 1993 using a complete intracorporeal technique with a purse-string suture. Modifications since then using extracorporeal techniques preformed under laparoscopic vision were first described in the early 2000s by William Spurbeck, Thom Lobe, and others and there is now a range of alternatives available.

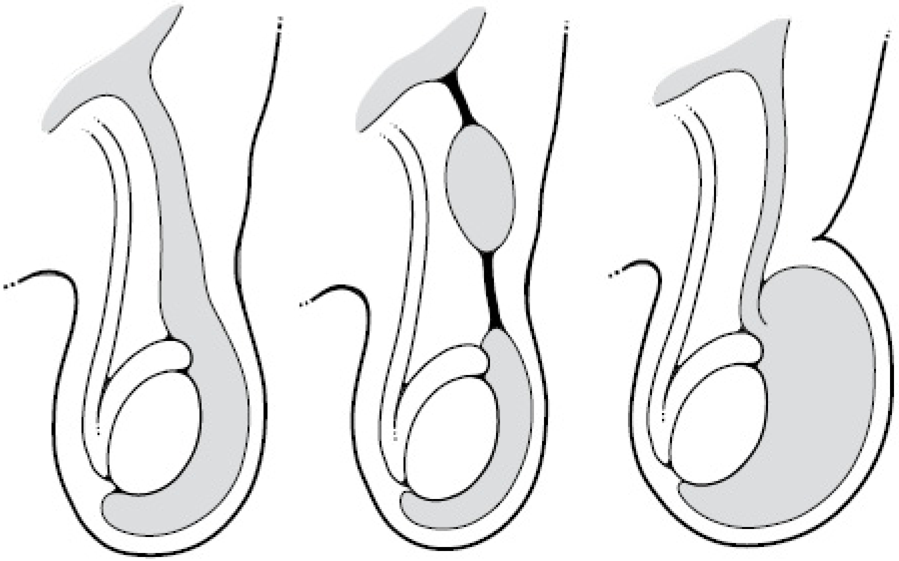
Today's surgeon now has the choice of open or laparoscopic repair of inguinal hernias from infancy onward.

PRINCIPLES AND JUSTIFICATION

In boys, congenital inguinal hernias are related to descent of the testis through the layers of the abdominal wall (**Figure 24.1**). The gonad starts intra-abdominally at the level of the kidney and reaches the internal ring at about 15–20 weeks' gestation primarily under the influence of insulin-like hormone 3 (INSL3) produced by testicular Leydig cells. Thereafter, led by the shortening gubernaculum, it passes through the internal ring under the influence of local calcitonin gene-related peptide (CGRP) released from the genitofemoral nerve and under the influence of testosterone. It drags with it a diverticulum of peritoneum on its anteromedial surface referred to as the "processus vaginalis." This normally fuses in >90% of full-term infants, obliterating the entrance to the canal, but failure of obliteration may result in a variety of inguinoscrotal anomalies, including an indirect inguinal hernia, a communicating hydrocele, hydrocele of the canal and a hydrocele of the tunica vaginalis. There are a number of obvious predisposing factors, particularly prematurity, but others include anterior abdominal wall defects (gastroschisis, exomphalos, and cloacal exstrophy), and conditions characterized by excess intraperitoneal fluid (e.g. ascites and ventriculoperitoneal shunts).


Peritoneal cavity
Obliterated processus vaginalis
Vas
Tunica vaginalis

(a) Normal (b) Inguinal hernia



(c) Complete inguinal hernia (d) Hydrocele of cord (e) Communicating hydrocele

24.1a–e

Direct inguinal hernias are much less common in children than adults but may be found in the preterm infant, and those with a neurological or myopathic condition (e.g. spina bifida).

In girls, the analogous diverticulum of processus through the internal ring was first described by Anton Nuck in 1691 and is referred to as the canal of Nuck. Herniation down to the level of the labia majora and hydrocele formation may also occur, albeit with less frequency than boys (1 : 4).

Clinical presentation

The majority of inguinal hernias in infants and children are indirect hernias. Boys are more commonly affected than girls (9 : 1), with a right-sided predominance of about 60 : 40. About 15% are bilateral. The diagnosis is often made on the basis of a bulge and can be observed in the groin with crying, straining, or standing. Scrotal enlargement or change in size may be noted.

Examination should confirm these observations but, not uncommonly, the diagnosis may depend on visualization by the referring pediatrician or parents. A photograph on a digital camera can be useful to remove doubt. Hernia and hydroceles should be distinguishable. Most of the former can be reduced with gentle pressure. If not, this implies an incarcerated (or even obstructed) hernia, which will be tender and cause distress. Hydroceles are not painful and cause no distress or obstructive symptoms. One should also be able to “get above” a hydrocele arising from the scrotum and feel a normal spermatic cord. Fluid in a hydrocele sac should also be “transilluminable,” giving a characteristic red glow with a cold light source. If there is still genuine doubt, ultrasound may be considered.

Inguinal hernia is a high-risk hernia, particularly in early infancy, as it is frequently complicated by incarceration, occasionally leading to intestinal obstruction and strangulation. In young infants with undescended testes and associated hernia, the testis is sometimes at risk of torsion or atrophy caused by compression of the vascular supply by a hernia sac filled with bowel at the level of the internal inguinal ring. The incidence of incarceration is highest in infants, particularly the premature, where an incarceration rate of up to 30% has been reported.

Indications

Most hydroceles will undergo spontaneous involution at 6–12 months and, as long as they do not change in size, these can simply be observed. Later-onset hydroceles are a candidate for surgery as involution is rare. All other inguinoscrotal anomalies require surgical intervention. Delay may result in complications such as intestinal obstruction in both boys and girls. The latter can also present with a tender mass in the canal or labia due to incarcerated ovary and Fallopian tube. This, too, needs urgent exploration as there is a risk of torsion or ischemic damage to the ovary.

PREOPERATIVE ASSESSMENT AND PREPARATION

The operation is usually performed shortly after the diagnosis is made. Attempts to reduce an incarcerated hernia using sedation and manual reduction are successful in more than 80% of cases. An elective operation is then typically carried out within 24 hours of the reduction. If surgery is delayed re-incarceration invariably occurs. In those instances of hernias in small premature infants already hospitalized in the neonatal intensive care unit because of other illnesses, then elective repair is carried out just before discharge and/or a weight greater than 2.0 kg. For infants diagnosed after discharge from the hospital who required ventilator support or who experienced episodes of apnea and/or bradycardia in the neonatal period, elective repair may be delayed until 50 weeks of corrected conception age to optimize their clinical status. Although most infants and children can be managed in an ambulatory setting, infants with bronchopulmonary dysplasia, anemia, prematurity (<50 weeks of corrected conception age), or those who required ventilator support at the time of birth should be observed after operative repair in an extended observation (23 hours) center and monitored for episodes of apnea and/or bradycardia.

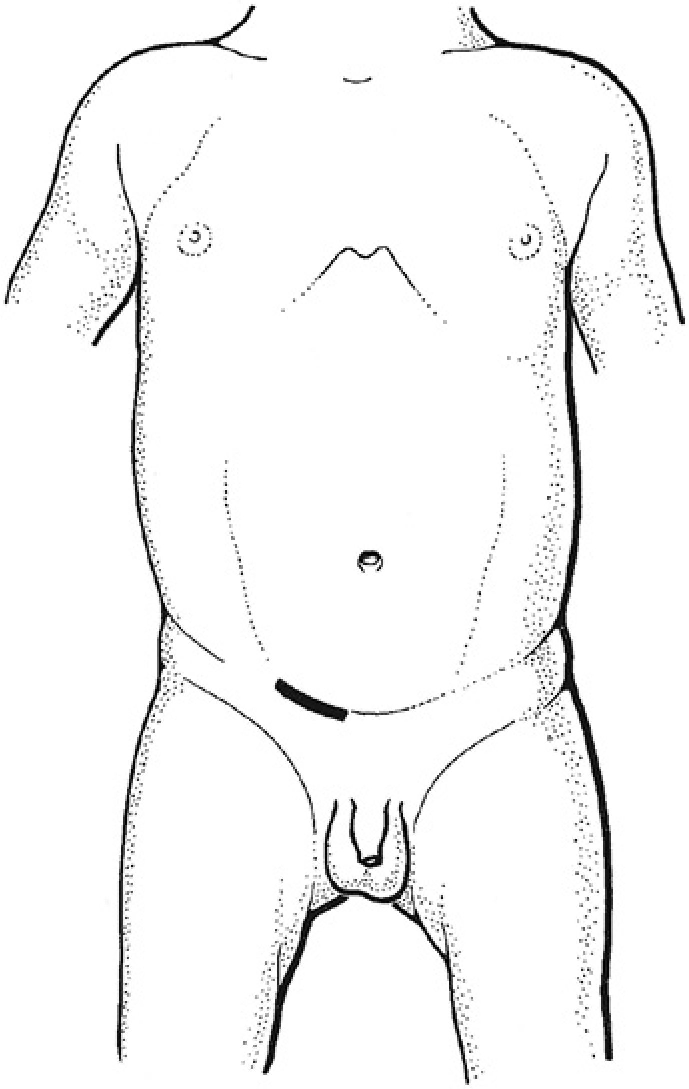
ANESTHESIA

The operation is usually performed under general anesthesia, although regional analgesia methods for inguinal hernia repair have attracted increasing interest. Caudal block, lumbar epidural block, ilioinguinal nerve block, and wound infiltration have all been used with varying success. Spinal anesthesia has become a popular technique, but some studies report intraoperative restlessness and pain during the repair, which may be due to peritoneal sac traction.

OPERATIONS

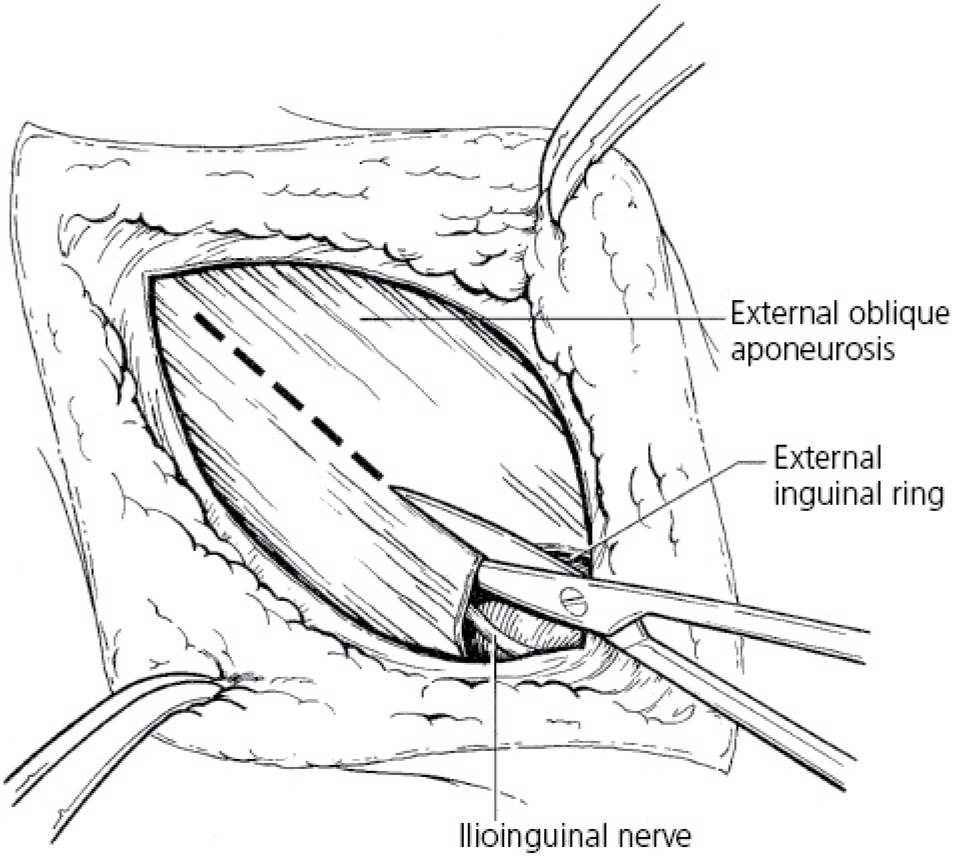
Open approach

A transverse incision is made in the lowest inguinal skin crease above the external inguinal ring on the affected side (**Figure 24.2**). Be aware of the superficial epigastric vein here to avoid bleeding. Scarpa's fascia is incised and the external oblique aponeurosis is identified. This is exposed and traced inferiorly to the inguinal ligament and medially to expose the decussating external (superficial) inguinal ring enveloping the emerging spermatic cord. This technique minimizes the risk of opening the inguinal canal too medially.



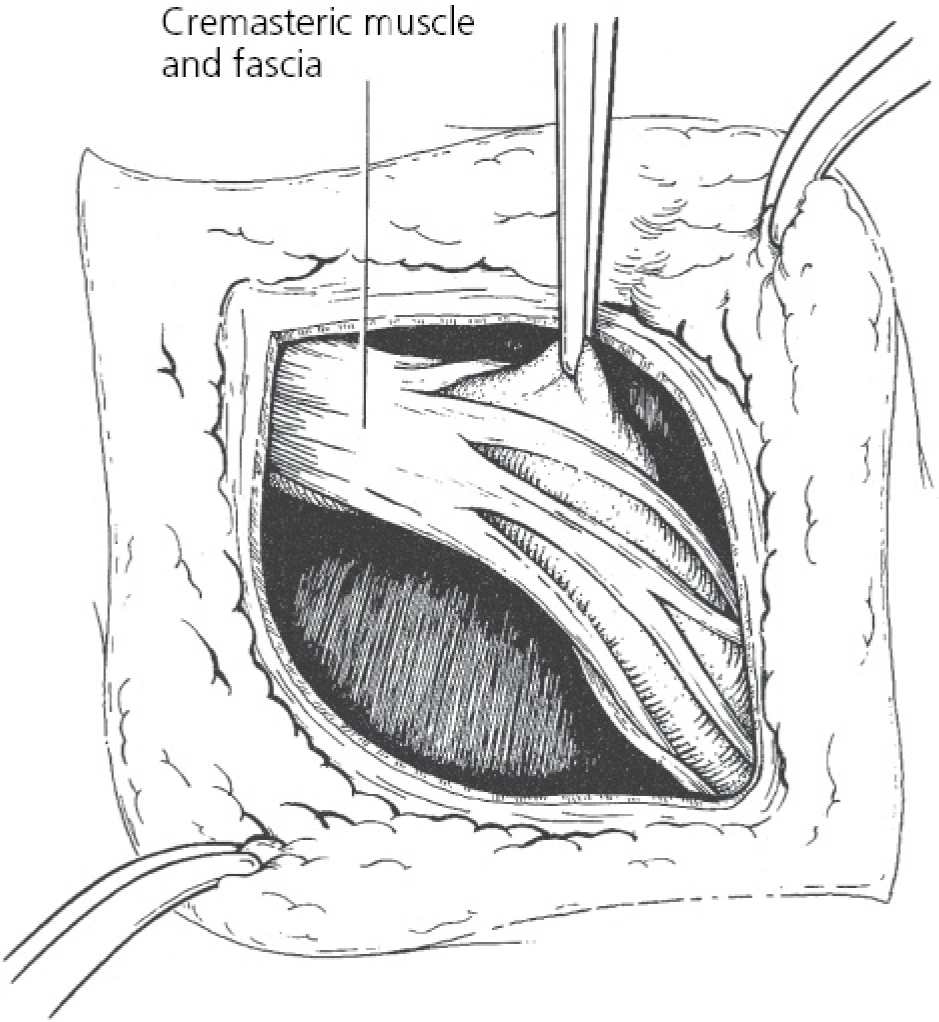
24.2

The external oblique aponeurosis is opened in the long axis of its fibers, for a distance of 1–2 cm near the deep inguinal ring. Care is taken during this maneuver to minimize accidental injury to the ilioinguinal nerve, lying adjacent to the cord and emerging through the superficial ring (**Figure 24.3**).


External oblique aponeurosis
External inguinal ring
Ilioinguinal nerve

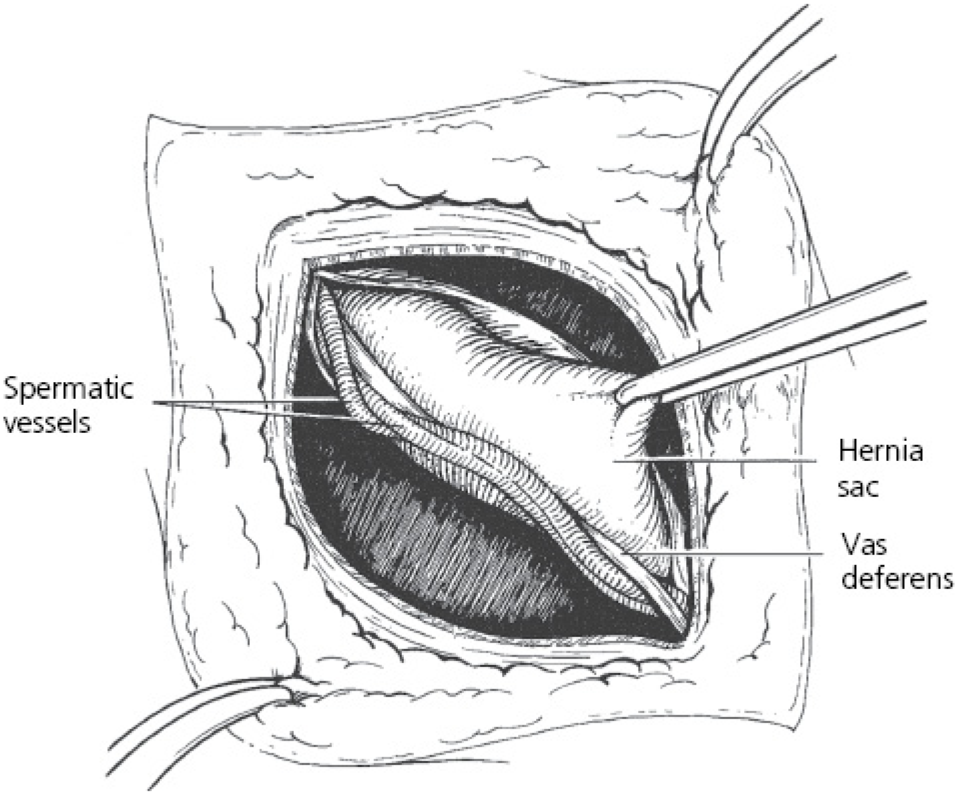
24.3

The undersurface of the superior leaflet of the external oblique is gently dissected free from the internal oblique and abdomen's transverse muscle. The inferior leaflet is mobilized down to the inguinal ligament. The iliohypogastric nerve lies at a higher level away from the cord. The layer of cremasteric muscle is teased open by blunt dissection onto the anteromedial surface of the cord and spread to expose the glistening peritoneum of the indirect hernia sac (**Figure 24.4**).



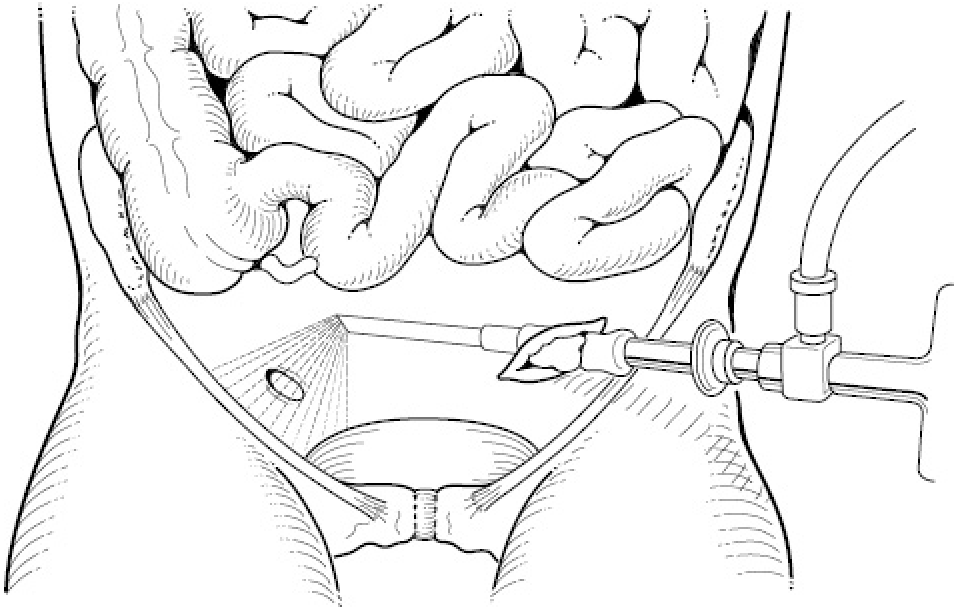
24.4

The sac is grasped with forceps and elevated anteromedially. Testicular vessels and vas deferens lie on the undersurface of the processus/hernial sac and are enveloped by it. These should be carefully identified and dissected free from the sac, which can be tricky, particularly in infants. Both vas and vessels are often intimately adherent to the sac and should be teased apart (**Figure 24.5**). Neither should be grasped directly with a forceps or a clamp as this can result in injury. The fundus of the hernia sac may be visible in the canal but more often extends toward the scrotum. Once the vas and vessels are identified, they are kept together using a silicone "vascular" sling. Enough is cleared laterally to divide the hernia sac between clamps. The proximal part is dissected toward the level of the internal (deep) inguinal ring, seeing retroperitoneal fat at the neck of the sac and/or the inferior epigastric vessels in its medial border.



24.5

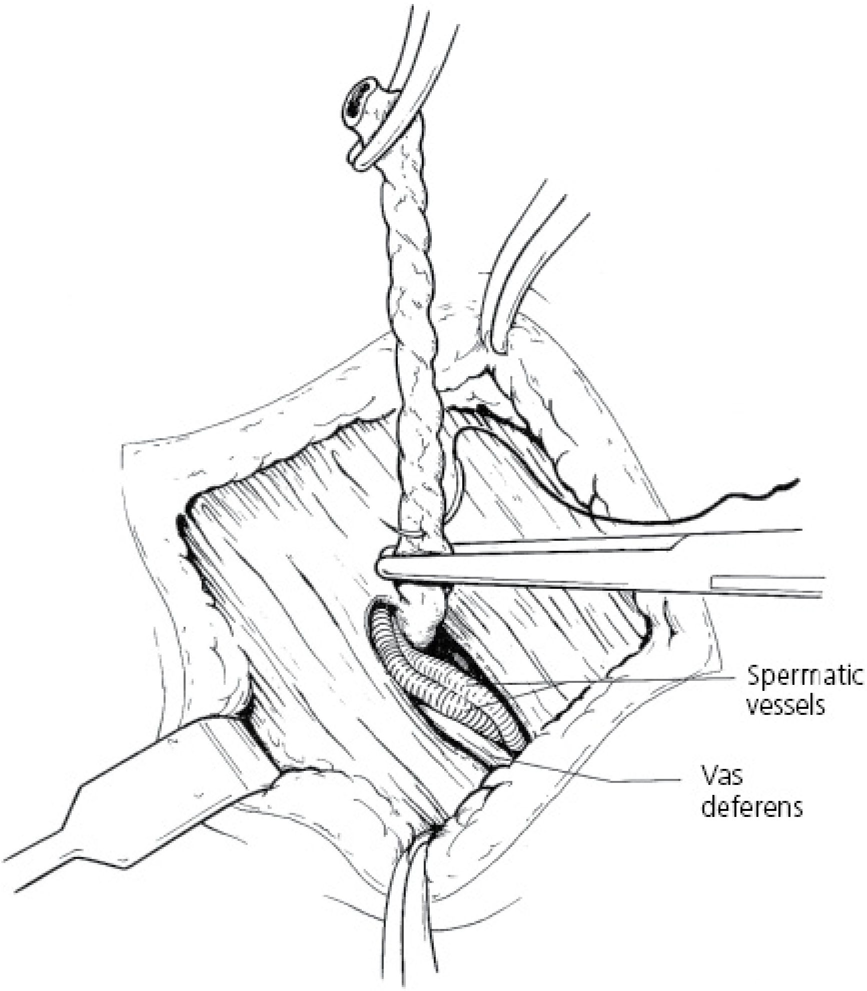
Laparoscopy through the inguinal hernia sac may be performed at this point to evaluate for a contralateral patent processus vaginalis (PPV). The sac can be opened on the most distal end and a 2 mm or 3 mm trocar is advanced into the abdomen and secured with a tie to minimize insufflation leak. The abdomen is insufflated with CO_2 to 8–10 mmHg pressure and a 70-degree laparoscope is advanced through the trocar into the abdomen to view the opposite internal ring (**Figure 24.6**). It may advantageous to have the patient in some degree of Trendelenburg position to aid in moving the bowel out of the pelvis and away from the contralateral internal ring.



24.6

The surgeon will need to decide whether to perform a contralateral repair if there is a contralateral PPV. If so, a similar repair as described above can be performed on the opposite groin. This technique avoids unnecessary *routine* contralateral exploration for all patients without a recognizable bulge on the opposite side and limits repair to those that have a demonstrable defect on laparoscopy. Still, it is important to note that not all patients with a PPV will develop into clinical hernias, potentially subjecting some to an unnecessary operation.

Following laparoscopy, the trocar is removed and the base of the sac twisted to ensure that all the contents are fully reduced (**Figure 24.7**). If there are contents and it is difficult to reduce, one should suspect a sliding component within the posterior wall of the sac. Sliding hernias often present as a more chronic hernia with the bulge often noted for a prolonged period of time prior to referral. The sac may contain bowel – partly intraperitoneal and partly extraperitoneal – or the bladder can be found medially. Alternatively, the cecum may come down on the right lateral side or the descending colon may be part of the sac on the left lateral side. Fallopian tube, ovary, and occasionally the uterus can be part of the sliding component in girls. These are all at risk of accidental injury. Failure to recognize a sliding component may also result in injury if the surgeon attempts to perform high suture ligation of the sac and includes the viscus in the suture. This component needs to be separated from the sac and may be facilitated by opening the sac and delicately dissecting off any intra-abdominal structures. High sac ligation at the internal ring can then be safely carried out. Forceps (traditionally a slotted spoon!) may then be placed at the base of the sac to protect the cord structures. The neck of the sac is transfixed twice with a non-absorbable 4/0 (or 3/0 in older children) suture ligation. Avoid a free tie because of the risk of dislodgment.



24.7

The distal end of the hernial sac is then opened on its anterior surface and excess tissue excised. If a separate hydrocele is present, this should be drained. If the internal ring is excessively large (usually defined as >1.5 cm), this can be reduced inferior to the cord with an interrupted 4/0 non-absorbable suture placed across the transversalis fascia vessels (*Marcy repair*). The floor of the inguinal canal is usually normal and requires no specific repair.

Sometimes there may be an associated direct inguinal hernia and definite posterior wall weakness. Again, this may be repaired by inserting a few non-absorbable sutures between the conjoint tendon and the inguinal ligament (*Bassini repair*).

The testis should be returned to a normal intrascrotal location at the end of the procedure. Administration of a local anesthetic (e.g. bipivacaine 0.25%, use 0.25 mL/kg per hernia repair side for a maximum of 3 mg/kg dose) along the ilioinguinal and iliohypogastric nerves will reduce postoperative pain.

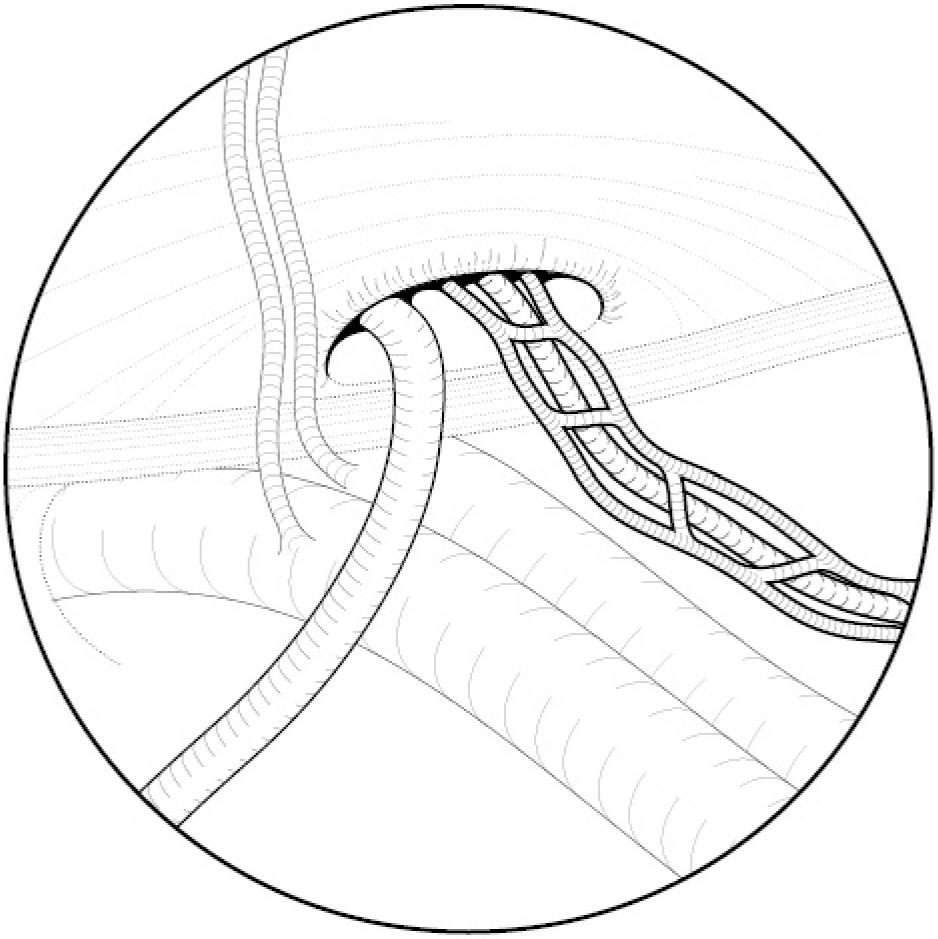
Wound closure is accomplished with an interrupted or running absorbable 4/0 suture approximating the previously mobilized external oblique leaflets to the external ring. Scarpa's fascia is closed with one or two interrupted 4/0 absorbable sutures. The skin edges are opposed with running subcuticular absorbable sutures or even tissue glue.

Laparoscopic approach

Laparoscopic hernia repair is an effective and increasingly popular alternative to open herniotomy, but there are multiple variations, including both intracorporeal and extracorporeal techniques.

The patient is placed in the Trendelenburg position. The surgeon and assistant stand at the head of the operating table, and the monitor is placed at the foot of the table. A 3 mm trocar is placed through an umbilical incision and the abdomen is insufflated with CO_2 to 5–8 mmHg pressure. Under telescopic vision, two 2 mm ports are placed superior and medial to the anterior superior iliac spine.

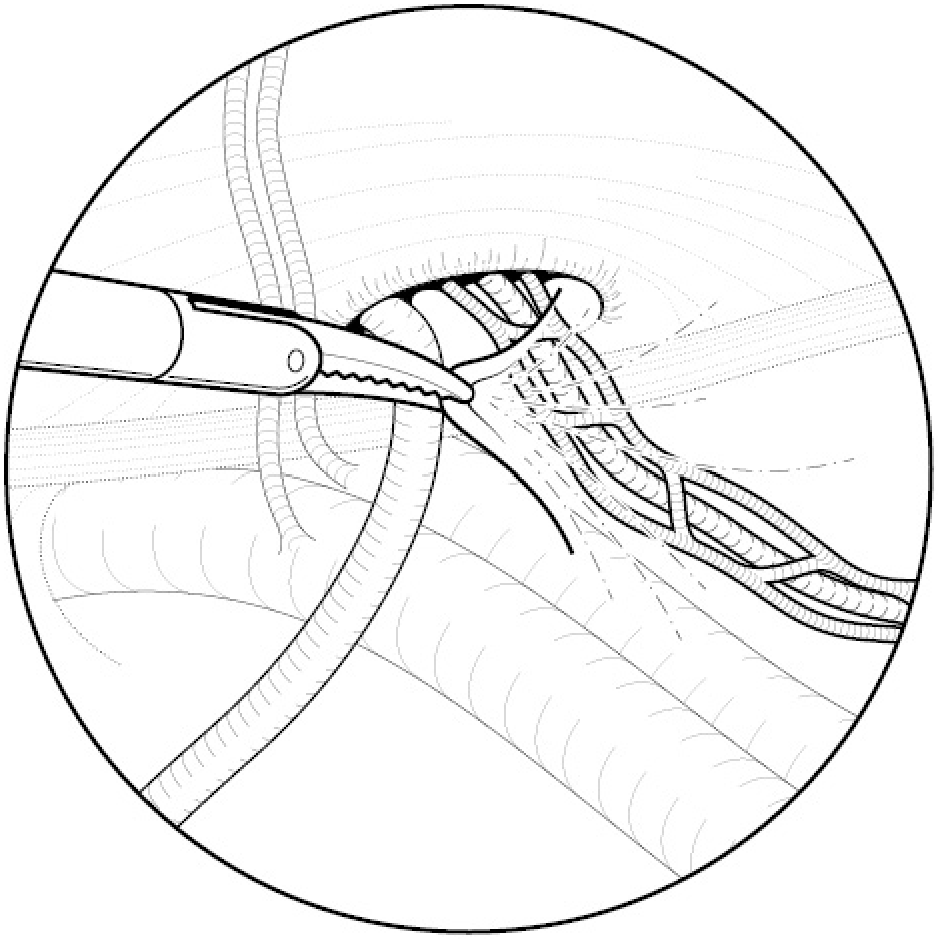
The internal opening of the hernia is confirmed, and the contralateral side is examined (**Figure 24.8**). A contralateral PPV is found in up to 40% of cases. As discussed above, it is controversial as to whether these should be closed during the same operation as not all will develop into a clinical hernia in the future, subjecting some patients to unnecessary operative risk. The authors generally counsel the parents about this during the consent process, and most parents request the contralateral PPV be repaired if found at the time of the operation.



24.8

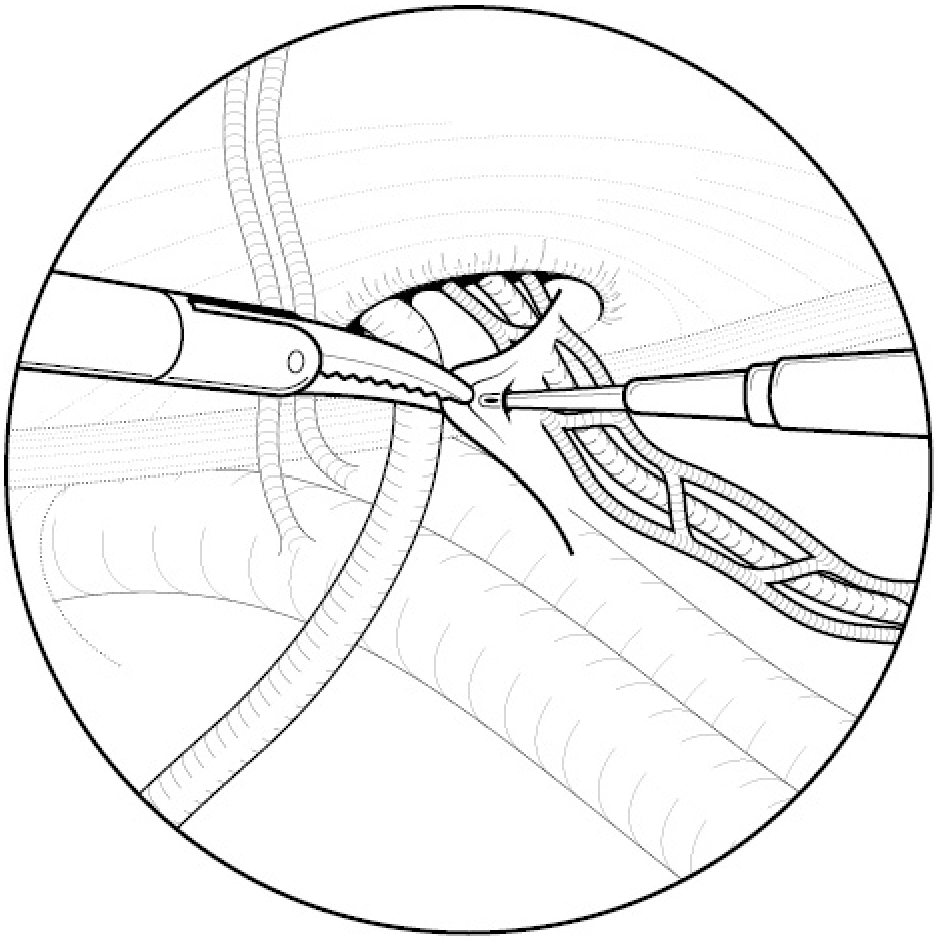
INTRAPERITONEAL PURSE-STRING CLOSURE

For boys, subperitoneal injection of normal saline to separate the vas deferens and testicular vessels from the peritoneum allows the safe application of a purse-string stitch (**Figure 24.9**). The peritoneum overlying the testicular vessels and vas deferens is picked up with a laparoscopic grasper.



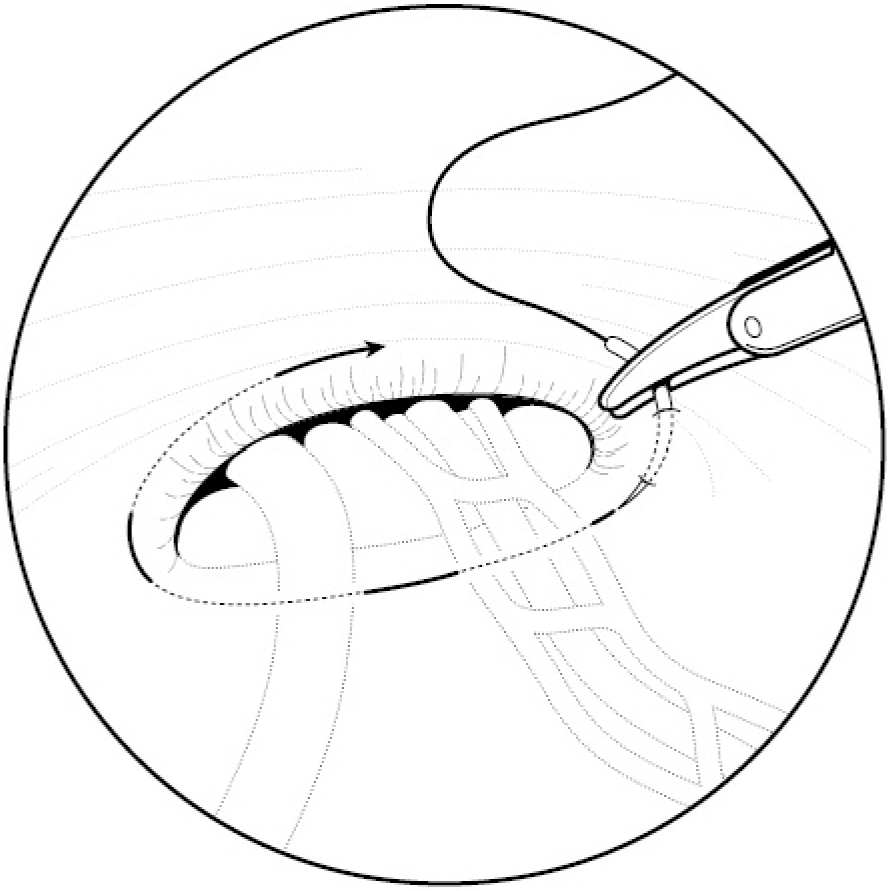
24.9

A needle is introduced and enters the space beneath the "tented" peritoneum under vision (**Figure 24.10**); either normal saline or local anesthetic is used to perform hydrodissection and 2 mL of normal saline are injected to elevate the peritoneum away from the vas deferens and testicular vessels. This should be performed first by injecting laterally around the ring, and then medially to ensure adequate separation of the vas and testicular vessels from the peritoneum. The injector is then withdrawn. Some surgeons also diathermize or incise the neck of the hernia sac with a diathermy hook. This has been shown in a rabbit model to create a more durable repair.



24.10

A 10–15 cm 4/0 non-absorbable monofilament (e.g. Prolene□) suture is passed into the peritoneal cavity through the abdominal wall next to the lateral port under vision. The purse-string stitch commences at the 2 o’clock position of the internal hernia opening (**Figure 24.11**); the needle should be seen to traverse the subperitoneal space free of the vas and testicular vessels at each successive “bite” of the peritoneum around the hernia ring.



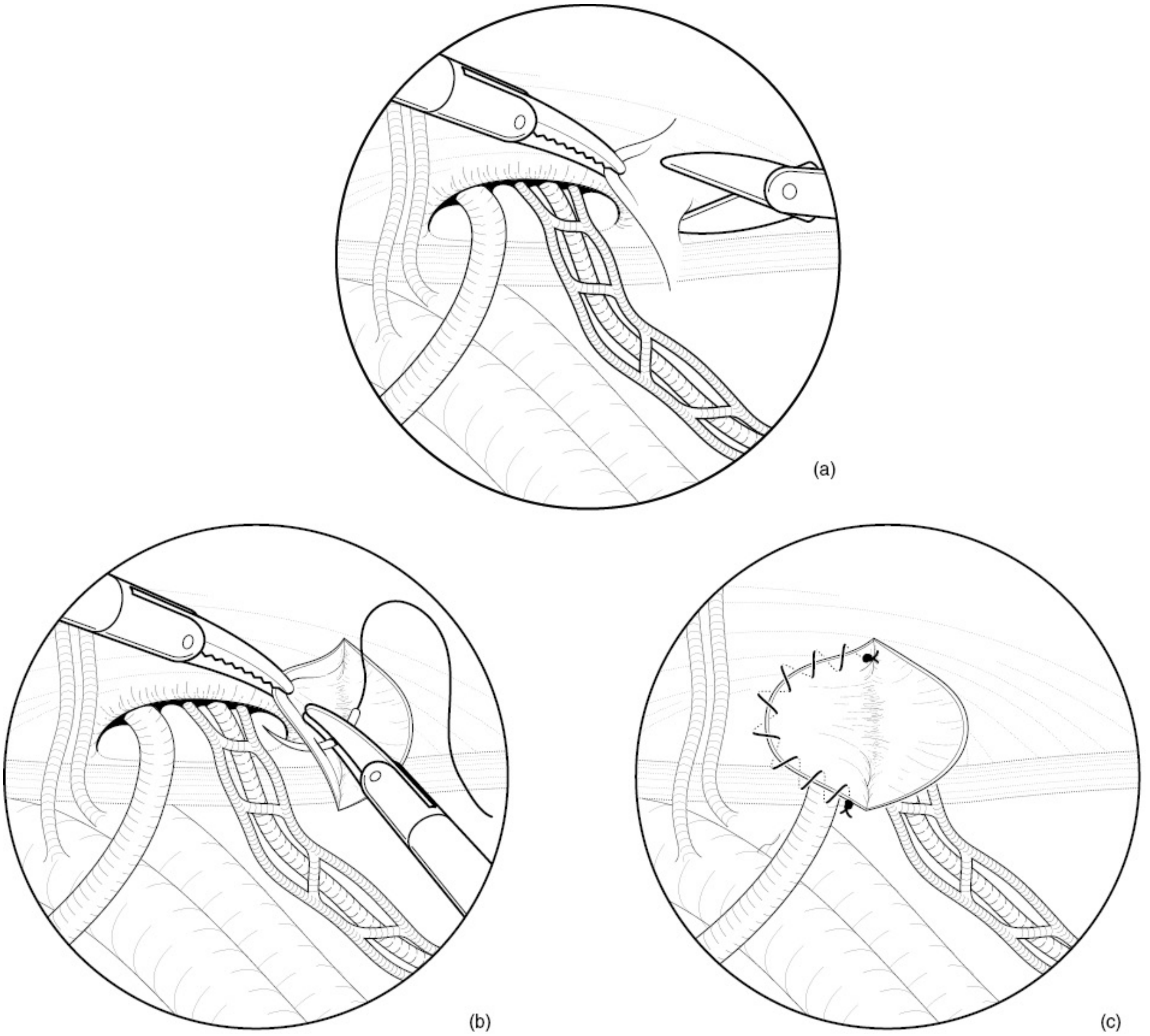
24.11

Care is taken to ensure that a "complete ring" of peritoneum has been included in the purse-string stitch without significant gaps; the exit and entry sites of successive "bites" of peritoneum should be as close as possible. For the anterior aspect of the internal hernia ring, sometimes it may be easier to pick up the peritoneum at the rim and apply "over and over" suturing until the 2 o'clock position is reached.

Before the stitch is tied, the intraperitoneal pressure is lowered to 2–4 mmHg, and gas in the hernia sac is expelled by compression of the groin/scrotum. The purse-string suture is tied tightly with several intraperitoneal knots. An airtight hernia closure is tested by increasing the intraperitoneal pressure transiently to 15 mmHg. There should be no gas leakage into the groin/scrotal hernia sac and the intraperitoneal pressure can be returned to 5–8 mmHg; if there is gas leakage, a second purse-string stitch can be added proximal to the first one. The ends of the stitch are cut, and the needle is retrieved through the abdominal wall. The ports are removed after release of the pneumoperitoneum. The umbilical wound is repaired in layers using absorbable sutures for fascia and skin, respectively. The lateral wounds are closed simply with Steristrips□ or tissue glue.

Alternative technique

The "flip-flap" technique involves a cut made on the peritoneum 1 cm lateral to the internal inguinal ring (**Figure 24.12a**). A flap of peritoneum is raised and "flipped" across to the internal inguinal ring to cover the hernia opening in the manner of a trap door (**Figure 24.12b**). Closure of the hernia ring is secured with a continuous suture between the free edge of the peritoneal flap and the adjacent hernia ring (**Figure 24.12c**). The use of a peritoneal flap avoids tension in the closure of large hernias.

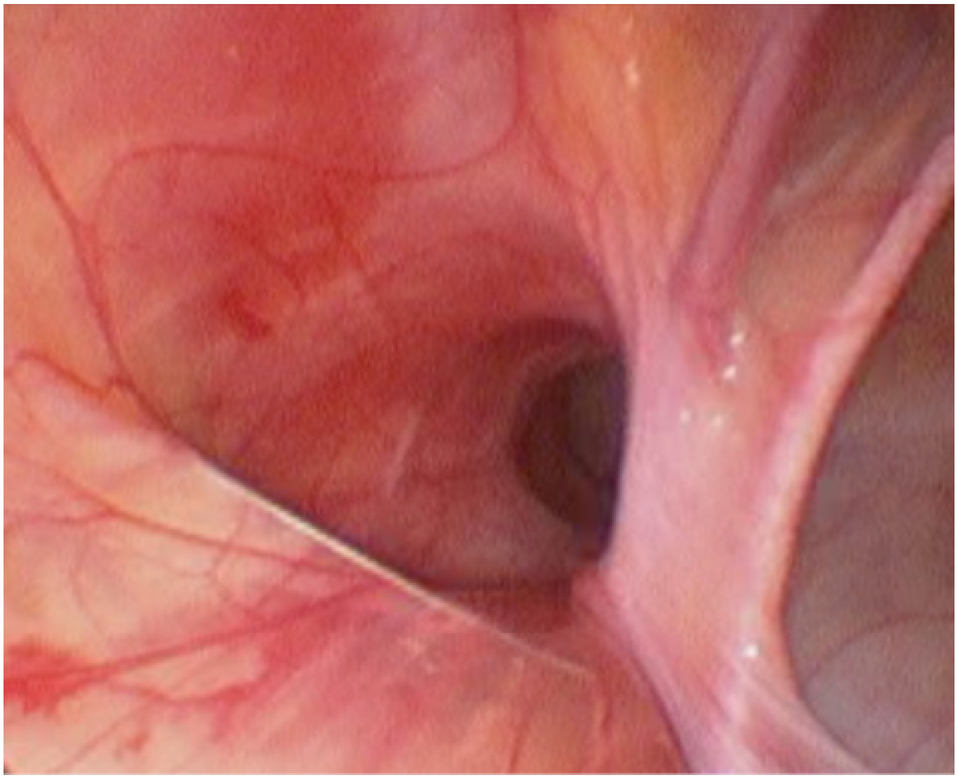


24.12a–c

EXTRAPERITONEAL CLOSURE

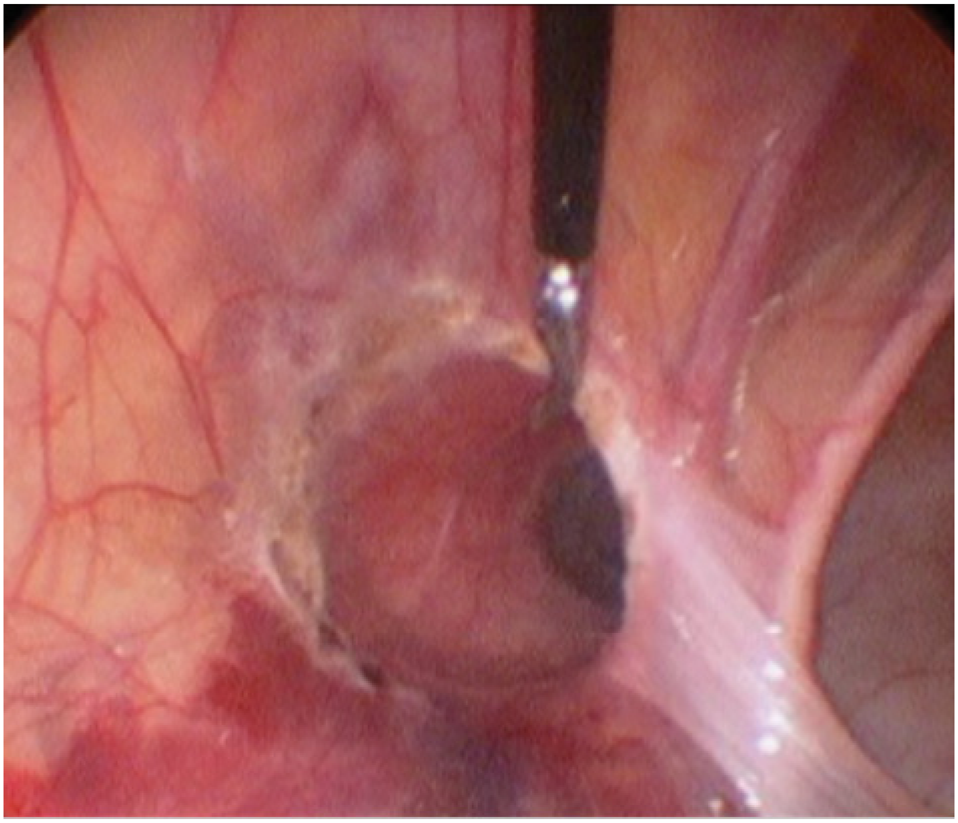
Laparoscopic closure of inguinal hernia can also be achieved by a two-port extraperitoneal encircling suture. A 3 mm telescope is introduced into the peritoneal cavity via an umbilical incision and pneumoperitoneum is established. A 2 mm or 3 mm grasper is inserted in the lower abdomen midway between the umbilicus and suprapubic tubercle under telescopic guidance through a stab incision. This port can be placed on the ipsilateral side of the hernia; however, the authors always place it in the left lower abdomen.

A small stab wound is made just at the 12 o'clock position of the internal inguinal ring lateral to the internal inguinal ring. Hydrodissection is again performed first laterally and then medially using a 25-gauge finder needle through this stab incision, injecting either normal saline or local anesthetic to separate the peritoneum from the vas deferens and testicular vessels. The incision is deepened to the preperitoneal space (**Figure 24.13**).



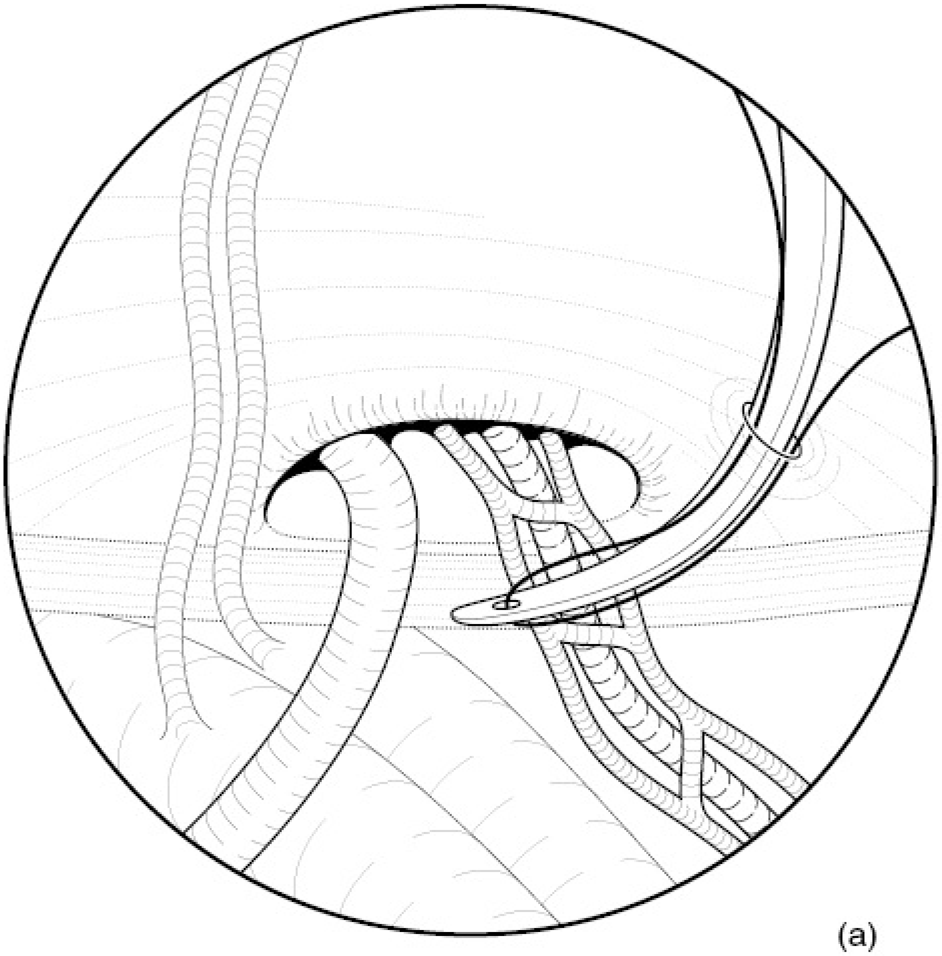
24.13

Thermal energy, through a stab incision, may be performed at this point around the internal ring to assist in creation of scar tissue and increase the durability of the repair (**Figure 24.14**).



24.14

There are many devices that may then be used to pass 3/0 non-absorbable, monofilament suture (e.g. Prolene□) around the internal ring, such as a hernia hook (Karl Storz, 26167HH, Tuttlingen, Germany; or its equivalent) or a spinal needle, which is the authors' preferred technique (**Figure 24.15a and b**).



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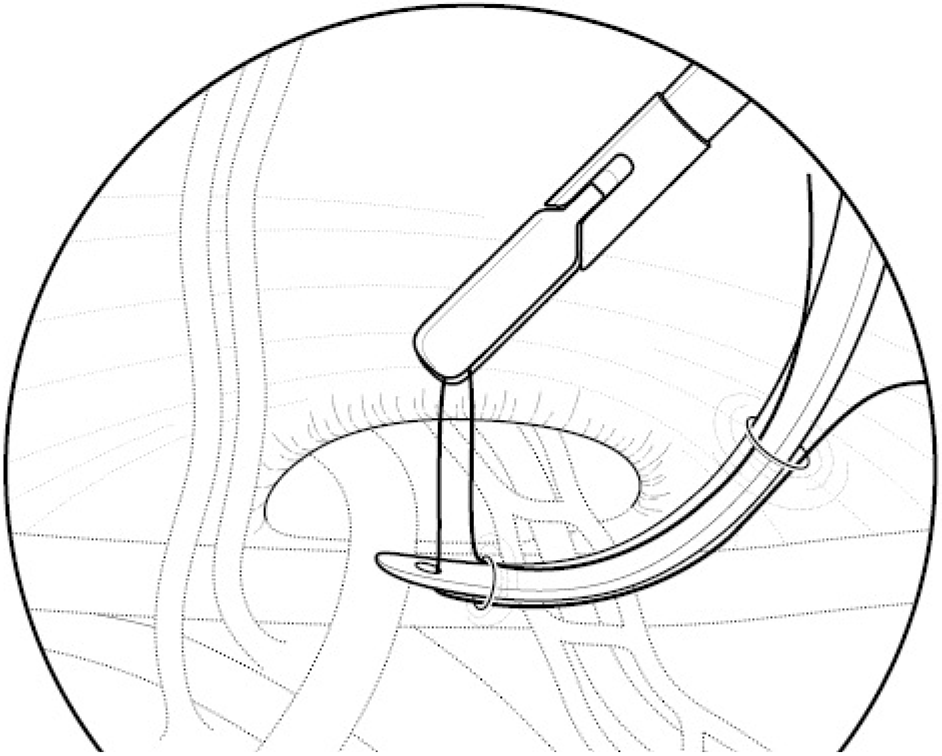
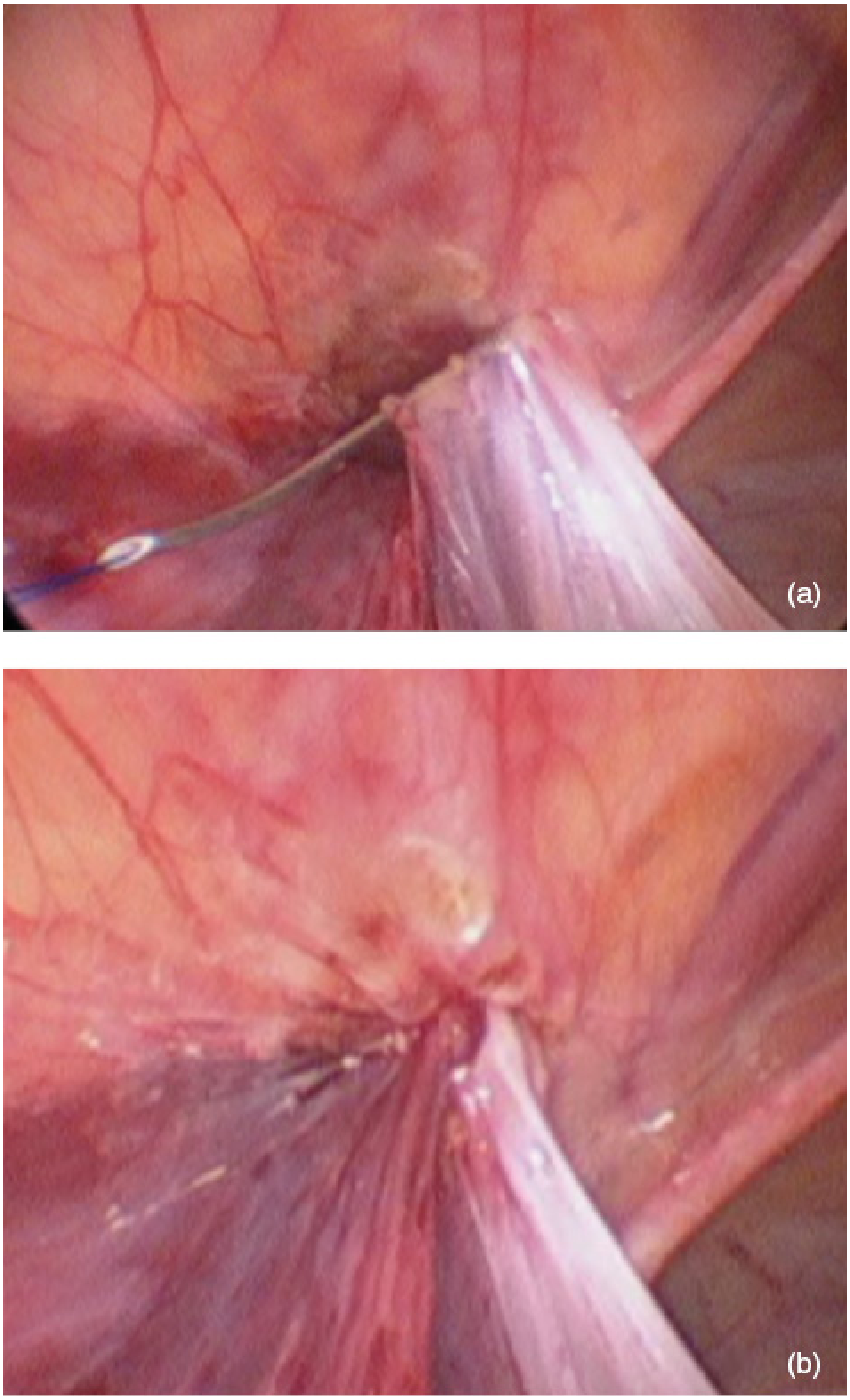


24.15a,b

The spinal needle is placed through the stab incision at the 12 o'clock position and passed laterally around the internal ring in the hydrodissection plane, over the cord structures. A Maryland dissector may be used to assist in lifting the peritoneum off the cord structures while passing the spinal needle. After the spinal needle has passed over the cord structures, the spinal needle should be pushed through the peritoneum into the abdominal cavity (**Figure 24.16a**). The looped monofilament suture is then pushed out partially from the needle, and the needle is removed, leaving the suture in place.



24.16a,b

The hook is withdrawn and reinserted into the anteromedial aspect of the preperitoneal space; the medial semicircle of the internal inguinal ring is similarly dissected as before until the previous hook entry site in the peritoneum is reached. The hook re-enters the peritoneal cavity. The suture is threaded through the eye of the hook. On withdrawal of the hook, the suture will completely encircle the internal inguinal ring. The distal hernia sac is compressed and pneumoperitoneum is released. The encircling suture is tied extracorporally and airtightness of the hernia closure is tested by reintroduction of pneumoperitoneum (**Figure 24.16b**).

POSTOPERATIVE CARE

With the exception of infants who require extended observation, most can be discharged within 2 hours of operative repair. There are no activity restrictions for infants, but older children should refrain from bicycle riding or other vigorous physical activity until their pain has subsided.

OUTCOME

Injury to the testicular vessels or vas deferens is unusual but may occur in approximately 3 per 1000 cases. If the vas deferens is divided, it should be repaired with interrupted 7/0 or 8/0 monofilament sutures. The use of magnifying loupes or an operating microscope will make the repair more precise.

Intraoperative bleeding is also an unusual complication unless the floor of the canal is weakened and requires repair. Needle-hole injury to the epigastric vessels or the femoral vein can usually be controlled by withdrawal of the suture and direct pressure.

Postoperative complications include wound infection, scrotal hematoma, postoperative hydrocele, and recurrent inguinal hernia. The wound infection rate at most major pediatric centers is quite low (~2%). An increased incidence of infection might be expected in patients presenting with an incarcerated hernia.

A recurrent inguinal hernia is a relatively uncommon complication in children with recurrence rates of <1% reported by experienced pediatric surgeons. Of these, most are noted within the first postoperative year. The major causes of recurrent inguinal hernia in children include:

- Missed hernial sac or unrecognized peritoneal tear
- Broken suture ligature at the neck of the sac
- Failure to repair (snug) a large internal inguinal ring
- Injury to the floor of the inguinal canal, resulting in a direct inguinal hernia
- Severe infection
- Increased intra-abdominal pressure (e.g. ventricular–peritoneal shunts and continuous ambulatory peritoneal dialysis)
- Cystic fibrosis and chronic cough
- Connective tissue disorders (e.g. Ehlers–Danlos syndrome)

Most surgeons would approach a first-time recurrent inguinal hernia repair through the previous inguinal site. However, a preperitoneal repair (*Cheatle-Henry type*) or traditional laparoscopic repair should be considered in selected cases with multiple recurrences.

Postoperative hydrocele may rarely occur after high ligation of the proximal hernial sac and incomplete excision of the distal portion. To avoid this complication, the anterior and lateral aspects of the sac are partially resected. The postoperative hydrocele often resolves spontaneously. Rarely, long-term persistence of the hydrocele may require a hydrocele aspiration and possible formal excision of the redundant hydrocele sac.

Testicular atrophy has been observed after repair of incarcerated hernias and in instances of large, acute tense hydroceles in young infants, but this is very rare after a typical hernia repair. There continues to be debate on the pros and cons of open versus laparoscopic inguinal hernia repair, as well as to the superiority of which method of laparoscopic repair. Several studies, including a prospective randomized controlled study, suggest that laparoscopic repair is associated with less pain, faster recovery, and improved cosmesis; in addition, laparoscopic hernioplasty allows detection and concomitant repair of contralateral hernia. With technical refinements of laparoscopic hernioplasty, the recurrence rate, which was high in some early series, is no longer a concern. While data on the long-term outcome of laparoscopic hernioplasty remain scarce, we have observed satisfactory results on patients with laparoscopic repair with more than 10 years' follow up.

FEMORAL HERNIA

PRINCIPLES AND JUSTIFICATION

A femoral hernia is a protrusion of preperitoneal fat or viscus through a defect in the femoral canal. It is not a common hernia in children but must be considered with a swelling in the inguinal region with the bulge inferior to the inguinal ligament. Femoral hernia may occasionally be confused with an enlarged swollen infected lymph node near the saphenofemoral junction (lymph node of Cloquet) just inferior to the inguinal ligament. Careful examination for a lower extremity focus of infection should be performed. Unfortunately, femoral hernias are often misdiagnosed and treated as inguinal hernias and therefore present following "recurrence."

Femoral hernias are most common in girls aged between 5 and 10 years. Some individuals advocate the use of laparoscopy in the child with presumed recurrent inguinal hernias because of the concern for a missed diagnosis. The anatomic boundaries of the femoral canal can be divided into anterior, posterior, lateral, and medial. The anterior border involves the iliopubic tract and/or the inguinal ligament. Posterior includes the pectineal ligament (Cooper's) and iliac fascia. The lateral boundary involves a connective tissue septum and the femoral vein. Medially, the canal is bordered by the aponeurotic insertion of the transversus abdominis muscle and tranversalis fascia.

Indications

There is no role for conservative management because of the risk of incarceration and strangulation. The fixed margins of the femoral ring result in early compression of swollen tissues and increase the risk of visceral compromise when incarceration occurs.

Choice of procedure

There are four possible methods to repair a femoral hernia:

- Low infrainguinal ligament repair (*Langenbeck*)
- Transinguinal Cooper's ligament repair (*McVay procedure*)
- Abdominal extraperitoneal repair (*McEvedy or Cheatle-Henry repair*)
- Laparoscopic repair

Although femoral hernia repairs have been performed using endoscopic techniques in adults, there are only limited reports in children and the authors have not used this approach. It may have a role in instances of recurrent hernias.

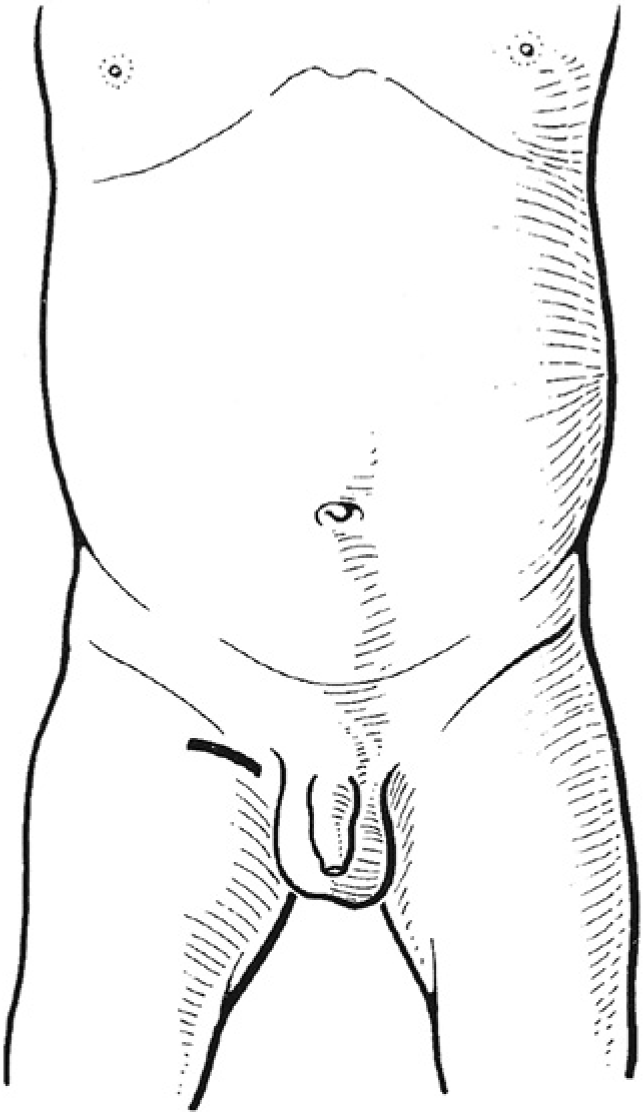
An inguinal or extraperitoneal approach should be used in instances of strangulated obstruction; however, since this is rarely seen in infants and young children, the low infrainguinal repair is preferred. If a concomitant inguinal hernia is present, an inguinal incision and McVay repair are recommended.

OPERATIONS

The low infrainguinal repair will be described here in detail as the other open approaches are dealt with in the section on inguinal hernia.

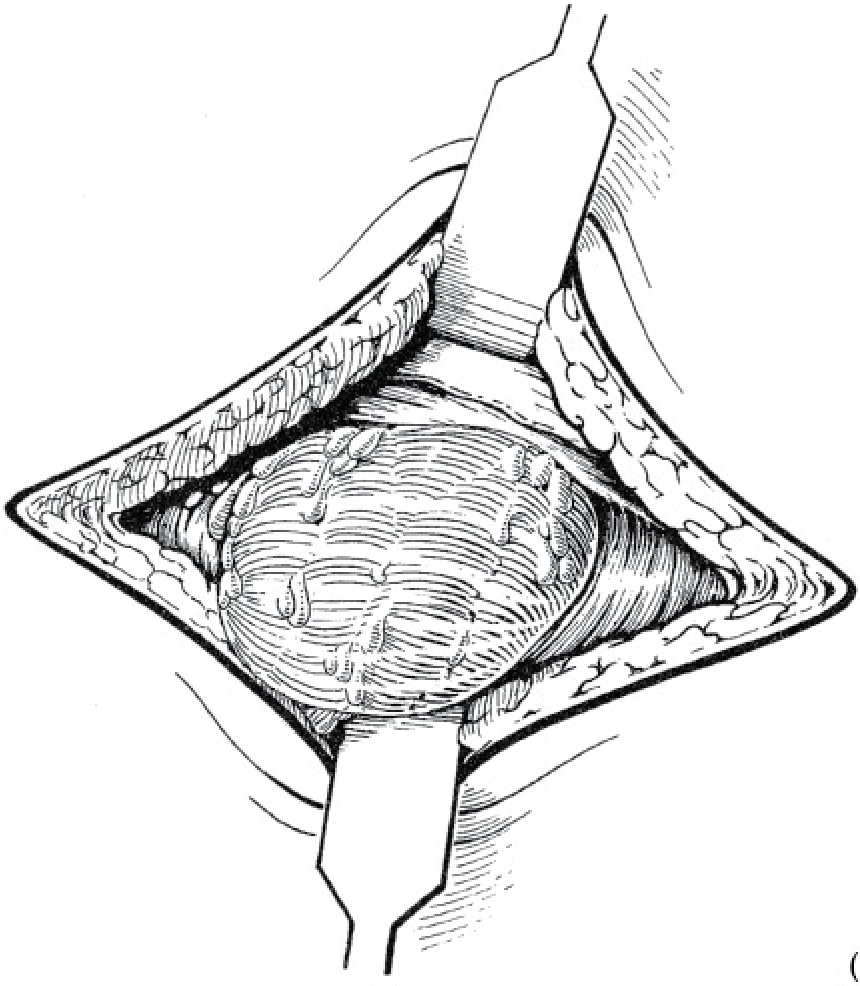
Low infrainguinal repair

A transverse incision is made in a skin crease over the mass from a point just inferior to the pubic tubercle medially, extending laterally just past the palpable pulsation of the femoral artery (**Figure 24.17**).

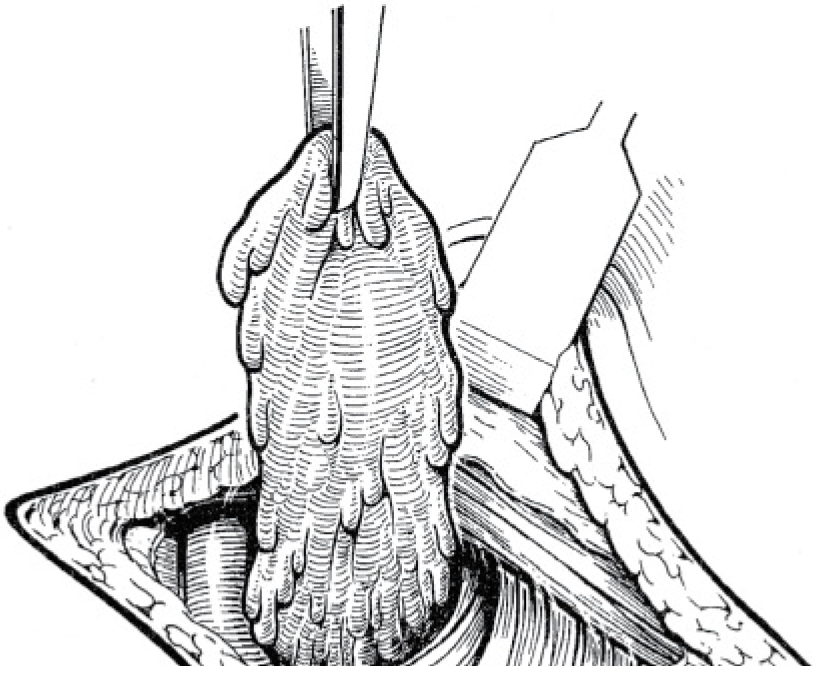


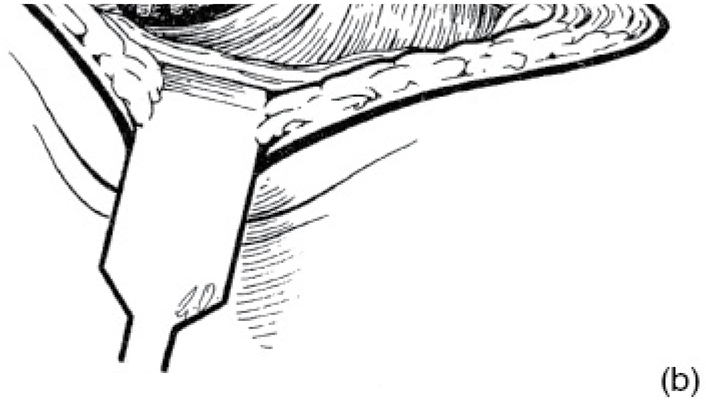
24.17

Hemostasis is effected with an electrocoagulator and the wound is deepened to expose the hernia sac bulge. Typically, the sac is covered by many layers (“onion skin”) including cribriform fascia and groin fat, and may overlie the femoral vein and extend upward over the inguinal ligament (**Figure 24.18a and b**).



(a)



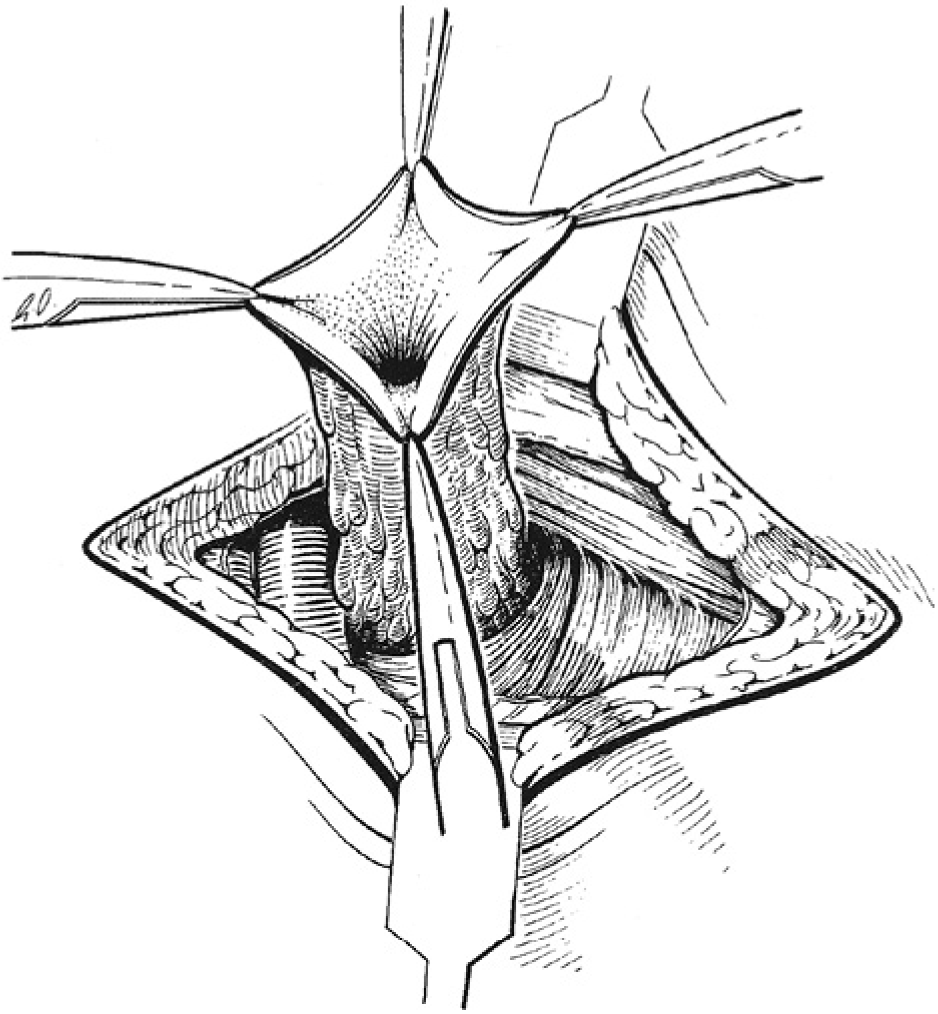


24.18a,b

All layers are incised, exposing the femoral hernia sac. Note the margins of the femoral canal as:

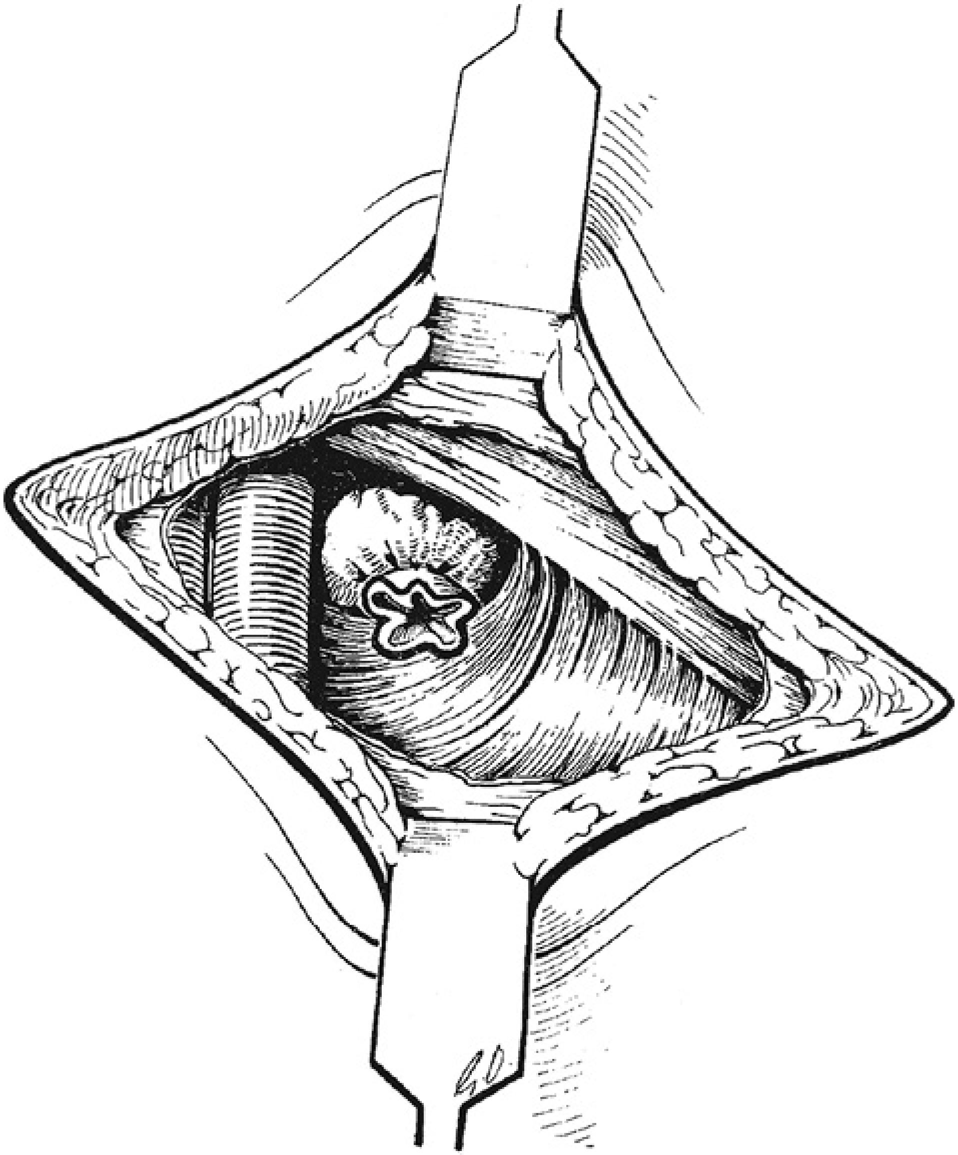
- Anterior – inguinal ligament (lower margin of external oblique aponeurosis)
- Posterior – pectineal ligament, fascia overlying pectineus
- Medial – lacunar ligament (of Astley Cooper)
- Lateral – femoral vein

The sac should be carefully palpated for visceral contents, which should be gently reduced through the defect (**Figure 24.19**). In the event that the sac is challenging to identify, an umbilical laparoscopic trocar can be placed and visualization of the femoral hernia can be noted and, with insufflation and guidance of the telescope, clear delineation of the sac can be accomplished. Occasionally, when incarceration is present and reduction is difficult, the sac can be released by carefully incising the insertion of the iliopubic tract into the lacunar ligament at the medial margin.



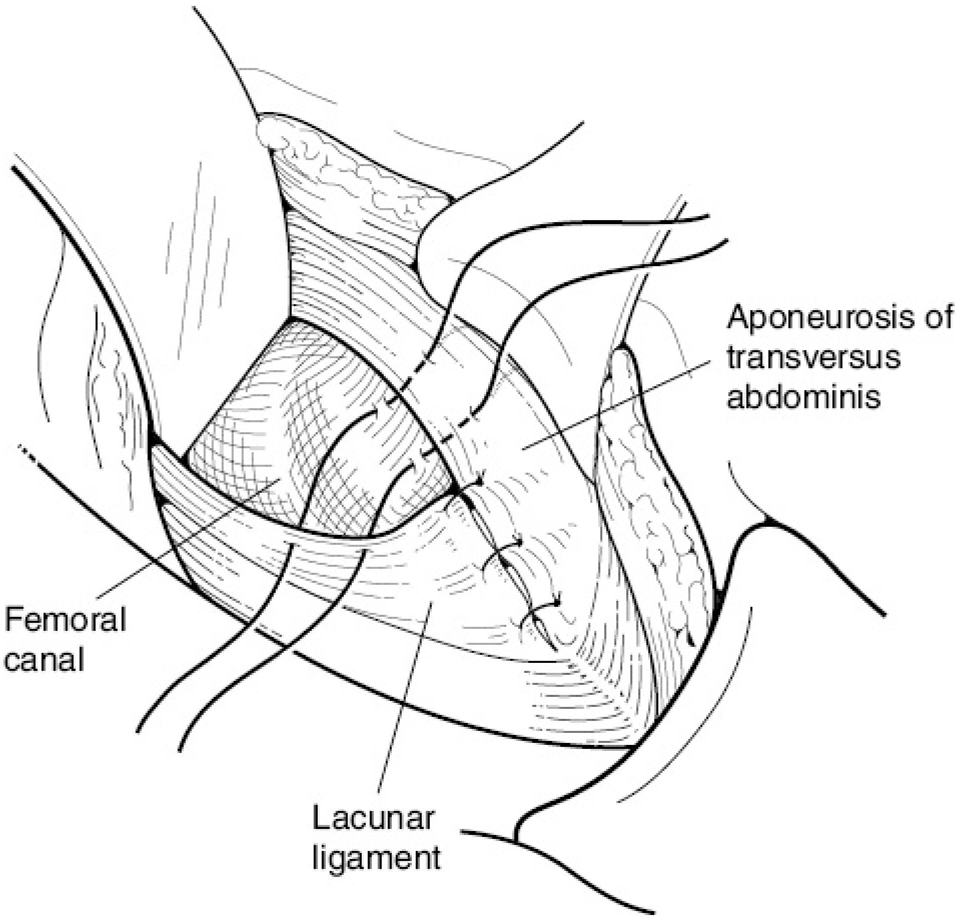
24.19

The peritoneal sac is small; however, it may have a considerable amount of retroperitoneal fatty tissue at its base. The sac can be opened (to ensure reduction of contents) if there is any question of a complete reduction. Hernial repair is initiated with high ligation of the sac similar to an inguinal hernia. This is accomplished with 3/0 non-absorbable suture ligatures. The sac may be bulky, however, and an alternative method of sac closure is inversion and reduction of the intact sac, placing one or two 3/0 non-absorbable purse-string sutures at a level just above the femoral defect (**Figure 24.20**).



24.20

Repair of the femoral defect is facilitated by placing a small retractor to raise the inguinal ligament superiorly in order to expose the pectineus fascia and lacunar ligament (**Figure 24.21**). If this is not performed, the femoral vessels may be traumatized with suture placement. The hernia repair is completed by suturing these two structures together, thereby obliterating the femoral canal medial to the femoral vein. All sutures are placed between the lacunar ligament and the inguinal ligament under direct vision before tying. The authors use interrupted 3/0 non-absorbable suture material in older children and 2/0 suture in adolescents. Special attention is given to avoid injury to the femoral vein either by the needle or by its compression when the sutures are tied. Gentle lateral retraction of the vein during suture insertion is useful.



24.21

Laparoscopic repair

During laparoscopic repair, the femoral hernia is seen as medial to the femoral vessels and below the inguinal ligament. Multiple variations of laparoscopic and laparoscopic-assisted repairs are described in both pediatric and adult literature, such as a laparoscopic patch-and-plug technique as well as closure of the femoral defect by suturing the iliopubic tract to the lacunar ligament intracorporeally. Some have described using laparoscopy to allow for a smaller incision of an open repair by inserting a laparoscope, identifying a femoral hernia, placing a Maryland dissector tip into the hernia, and then, on the outside, cutting on top of the Maryland tip to allow for a small incision directly on top of the defect.

OUTCOME

Wound infection may develop in ~1% of cases. It should be recognized promptly and the wound opened or administration of oral antibiotics should be initiated early to avoid possible extension of a closed infection to the deeper tissues. Wound hematoma is rarely observed and may be caused by an unrecognized tear in a saphenous/femoral venous branch.

Pain and ipsilateral leg swelling may be the result of compression of the iliac or femoral vein. This can be obviated or minimized with meticulous attention while placing the hernia repair sutures as previously described.

Since a femoral hernia is relatively rare in children, data concerning recurrence are not available and a recurrence rate of 1–2% in children might be expected. In the event of a recurrence, one must rule out an associated inguinal hernia. Some have adopted the preperitoneal approach for recurrent repair due to the higher rate of secondary recurrences. Since recurrences may be the result of a localized collagen defect and direct reapproximation of the weakened tissue may not be appropriate, the use of a prosthetic buttress may be indicated. The posterior approach will avoid reoperation through a previous surgical site, allow evaluation and exclusion of all groin-related hernia defects, facilitate the repair by easy visualization of lacunar ligament and the iliopubic tract, and, if a prosthetic buttress is chosen, the principles of distributing even intra-abdominal pressure to the prosthetic patch are employed.

UMBILICAL HERNIA

PRINCIPLES AND JUSTIFICATION

An umbilical hernia is very common in infants and young children. The hernial sac protrudes through a defect in the umbilical ring due to a failure of complete obliteration at the site where the fetal umbilical vessels (umbilical vein and the two umbilical arteries) enter the abdominal wall.

Approximately 20% of full-term neonates may have an incomplete closure of the umbilical ring at birth, rising to 80% of premature infants weighing <1.5 kg. Umbilical hernia is more common in girls than boys and black children have a much higher incidence than white children. The reason is obscure.

The hernia becomes more apparent during episodes of crying and straining and may result in considerable protrusion. It often causes considerable parental anxiety and frequent requests for operative repair in early infancy. Although rupture or incarceration of an umbilical hernia occurs, this is distinctly uncommon. It is rarely a cause of pain or other symptoms and as almost 80% of hernias will decrease in size and close spontaneously by 3–4 years of age, early age operation needs specific justification.

Indications

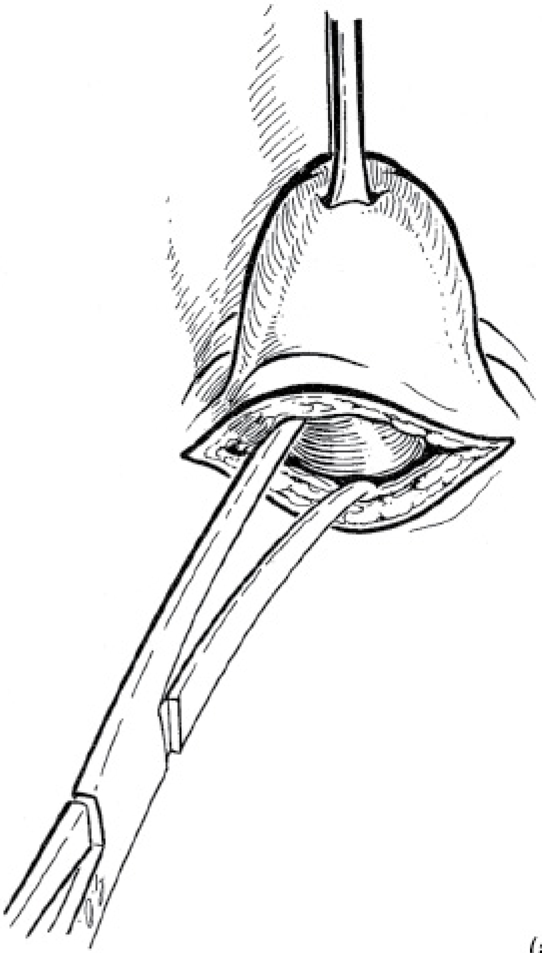
As the majority of these low-risk hernias will close spontaneously, it is safe to wait until the child is 3–5 years of age (particularly if the umbilical ring is <1.5 cm in diameter) before attempting repair. In contrast, defects of >2 cm diameter rarely close spontaneously. Hernia defects that do not close by 4 years of age should be electively repaired and, if the ring is considerably >2 cm or they have symptoms, it could even be done before this age.

Given the data on potential neurodevelopmental delay associated with anesthesia exposure in younger children, we choose to delay repair of asymptomatic umbilical hernias until after age 3.

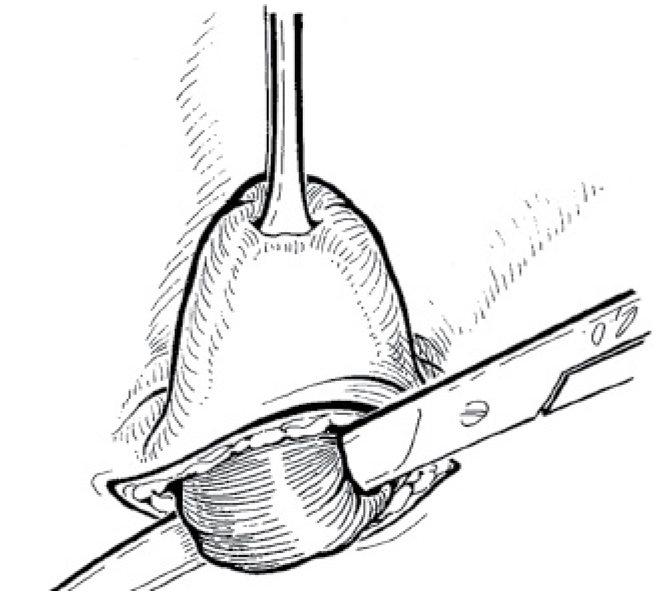
OPERATION

A curved (“smile”) incision is made in a natural skin crease immediately below the umbilicus. A supraumbilical incision is also acceptable, especially if there is an associated supraumbilical component. Placement of four-quadrant traction by the assistant on the abdominal wall and slight upward traction of the defect allows selection of the site of the incision. The curved incision should typically not extend beyond 180 degrees.

The subcutaneous tissue is incised and bleeding points controlled with a fine tip electrocoagulator. With upward traction on the inner margin of the upper lip of the incision, dissection is carried out down along the sac to the level of the anterior abdominal wall fascia (**Figure 24.22a**). By blunt dissection with a mosquito clamp, a plane is developed on either side of the sac, extending superiorly to gain control of the entire circumference of the sac (**Figure 24.22b**). Any contents in the sac should be reduced into the peritoneal cavity. If the sac is large, the surgeon or assistant places an index finger in the skin defect to evert the sac where it is attached to the skin. The sac is dissected free from its skin attachments, preserving the umbilical skin for an umbilicoplasty. Separation of the sac may require its transection near the skin to preserve the umbilicus for cosmetic purposes.



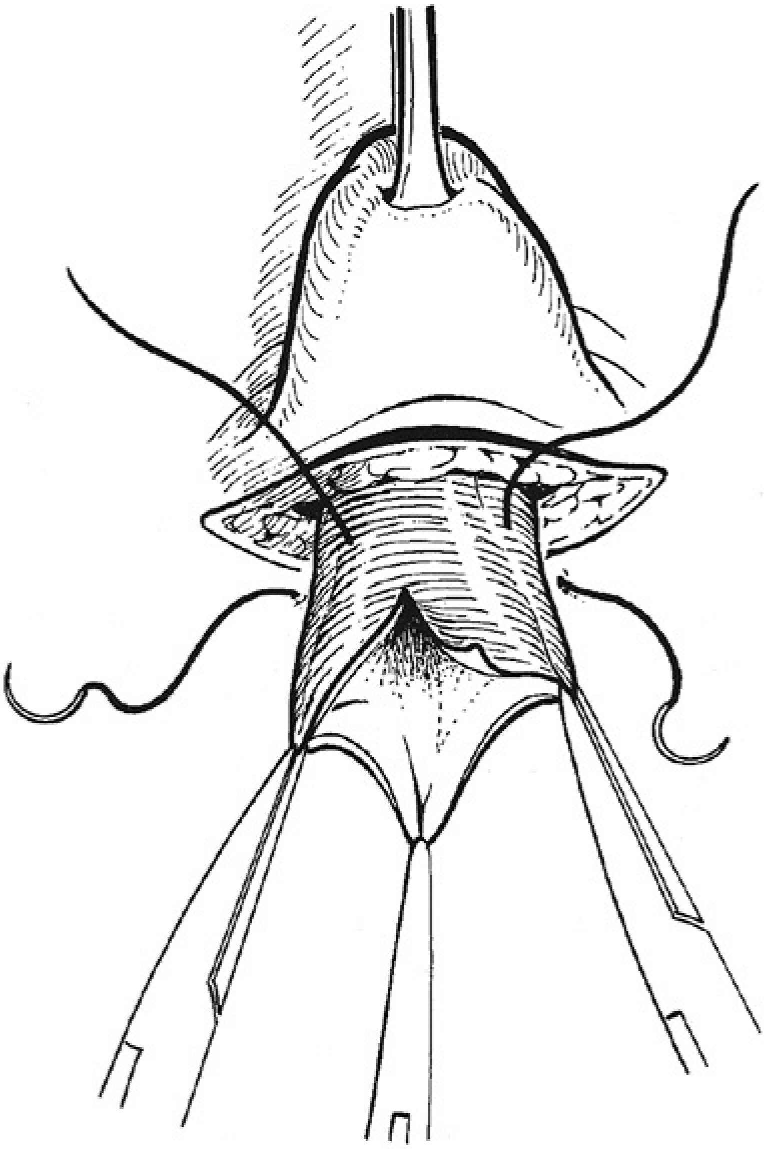
(a)



(b)

24.22a,b

The entire sac is elevated by mosquito clamps to maintain control of the edge of the defect and to have direct visualization during placement of sutures to avoid visceral injury. The sac is opened and any contents reduced. The rim of the defect is identified and the sac incised to allow placement of sutures starting at the corner farthest from the surgeon. Continuous or interrupted 3/0 (infants and young children) or 2/0 (older children and teenagers) sutures are placed (**Figure 24.23**). The sutures are elevated to maintain upward traction on the abdominal wall. The sac is partially excised at the level of the abdominal wall as more sutures are placed. A traction suture may also be placed at the corner of the transverse wound closest to the operating surgeon to offer exposure as the remaining sac is excised and sutures placed. If considerable redundant tissue is present or the initial tissue layer seems sparse, a layer of fascia can be imbricated over the initial line of repair with interrupted or continuous absorbable suture.



24.23

The benefit of performing an umbilicoplasty is controversial as it is unclear if it leads to a better long-term cosmetic outcome. For those who choose to perform an umbilicoplasty, it is performed by inverting the undersurface of the redundant umbilical skin to the anterior abdominal wall fascia with one or two interrupted 4/0 absorbable sutures. Any remnant of the peritoneum on the umbilical sac that is adherent to the skin may safely be left behind, excised, or cauterized. In children with a very large protuberant hernia with redundant skin, following the removal of the sac and fascial closure, the umbilicoplasty can sometimes be frustrating to the surgeon and patient. The volume and variety of literary material on the repair of umbilical defects attest to the fact that there exists no single ideal approach. The management of the excess umbilical skin can be completed using numerous methods, such as a purse-string suture, complicated V–Y advancement procedures, four equilateral triangular skin flaps, or the Mercedes-Benz umbilicoplasty (Video 24.1: Laparaoscopic hernia repair).

🎥

POSTOPERATIVE CARE

Postoperative activity restrictions are similar to those for an inguinal hernia repair.

OUTCOME

Complications are unusual and are limited to a wound infection (~1%) or an occasional wound hematoma. Bowel injury is the rarest of complications. Recurrence is rare as well, and those that do recur are typically in children with associated comorbidities, such as long-term continuous ambulatory peritoneal dialysis, ventricular peritoneal shunts, or a connective tissue disorder.

SUPRAUMBILICAL AND EPIGASTRIC HERNIAS

PRINCIPLES AND JUSTIFICATION

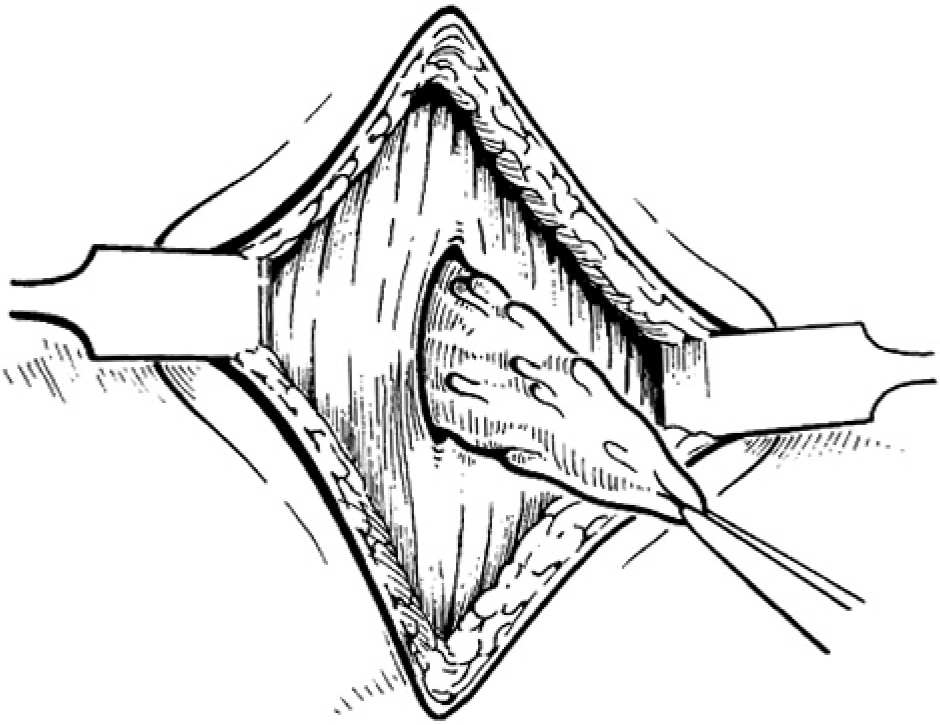
A number of defects may be observed along the linea alba between the xiphoid process and the umbilicus. Failure of fixation of the medial borders of the rectus abdominis muscles at the linea alba in infants results in a large, bulging defect (a diastasis or divarication of the recti) which is of virtually no consequence and resolves spontaneously as the baby grows and the linea alba becomes a firm structure.

Epigastric hernias usually occur in the mid-epigastrium. The actual defect may be small; however, it is often symptomatic. Fat from the falciform ligament or the omentum can incarcerate and cause pain. In some cases, a tender mass of incarcerated fatty tissue can be palpated in the defect. Since epigastric hernias do not spontaneously close and are often symptomatic, they should be repaired on an elective basis. If they are asymptomatic, some surgeons do not believe they should be repaired. As the hernial defect may be small, it is wise to mark the skin over the exact site prior to the surgical procedure with the child awake in a standing position.

OPERATION

A transverse incision is made directly over the pre-marked area identifying the site of the hernia defect or, in the case of a supraumbilical defect, a supraumbilical frown incision may be made to maximize cosmesis.

There is no peritoneal sac identified in most instances of epigastric hernia, simply a fatty protrusion through the linea alba (**Figure 24.24**). This may be suture ligated, excised, or inverted and reduced into the peritoneal cavity. The small defect (not uncommonly the size of a pencil tip) is repaired with interrupted 3/0 absorbable sutures.



24.24

ACKNOWLEDGMENTS

We thank the late Jay L. Grosfed, Scott A. Engum, and Paul K.H. Tam for the basic structure and text of this chapter taken from the seventh edition.

VIDEOS

OPS_8e_video_24.1 Laparoscopic extraperiotneal inguinal hernia
OPS_8e_video_24.2 Laparoscopic Ponsky inguinal hernia repair

FURTHER READING

Adibe OO, Hansen EN, Seifarth FG, et al. Laparoscopic-assisted repair of femoral hernias in children. *J Laparoendosc Adv Surg Tech A* 2009; 19: 691–4.

Blatnik JA, Harth KC, Krpata DM, et al. Stitch versus scar – evaluation of laparoscopic pediatric inguinal hernia repair: A pilot study in a rabbit model. *J Laparoendosc Adv Surg Tech A* 2012: 22: 848–51.

Chan KL, Chan HY, Tam PKH. Towards a near-zero recurrence rate in laparoscopic inguinal hernia repair for pediatric patients of all ages. *J Pediatr Surg* 2007; 42: 1993–7.

Chan KL, Hui WC, Tam PKH. Prospective, randomized, single-center, single-blind comparison of laparoscopic vs open repair of pediatric inguinal hernia. *Surg Endosc* 2005; 19: 927–32.

Kelly KB, Krpata DM, Blatnik JA, Ponsky TA. Suture choice matters in rabbit model of laparoscopic, preperitoneal, inguinal hernia repair. *J Laparoendosc Adv Surg Tech A* 2014; 24: 428–31.

Lee SL, DuBois JJ. Laparoscopic diagnosis and repair of pediatric femoral hernia: Initial experience of four cases. *Surg Endosc* 2000; 14: 1110–13.

Ostlie DJ, Ponsky TA. Technical options of the laparoscopic pediatric inguinal hernia repair. *J Laparoendosc Adv Surg Tech A* 2014; 24: 194–8.

Reyna TM, Hollis HW, Smith SB. Surgical management of proboscoid herniae. *J Pediatr Surg* 1987; 22: 911–12.

Schier F, Montupet P, Esposito C. Laparoscopic inguinal herniorrhaphy in children: A three-center experience with 933 repairs. *J Pediatr Surg* 2002; 37: 395–7.

Treef W, Schier F. Characteristics of laparoscopic inguinal hernia recurrences. *Pediatr Surg Int* 2009; 25: 149–52.

Yip KF, Tam PKH, Li MKW. Laparoscopic flip-flap hernioplasty: An innovative technique for pediatric hernia surgery. *Surg Endosc* 2004; 18: 1126–9.

25

Exomphalos/omphalocele

MARK DAVENPORT AND ANNA-MAY LONG

The terms exomphalos and omphalocele are interchangeable throughout most of the literature. Both are derived from the Greek word *omphalos* (ὀμφἄλός), which means "navel," "centre," or "hub." According to Greek legend, Zeus sent two eagles across the world to meet at its centre, the "navel" of the world. This was in Delphi, site of the oracle and a famous temple to Apollo, and was marked by placing an omphalos stone there.

HISTORY

Descriptions of umbilical herniation and its treatment dating from the 1st century BCE were written by Celsus in *De Medicina*. He, however, warned against the treatment of infants and "those which are excessively large." Amboise Paré (1510–1590) described the presence of bowel within the newborn umbilical cord but also cautioned against intervention; "Let not the Chirurgian assay to open the abscess, for if it be opened, the guts come out through the incision." Manual reduction of the extracorporeal bowel, accompanied by prolonged external compression was described by English surgeons such as Hamilton, Astley Cooper, and George Hey in the early 19th century accompanied by some successes.

A description of the successful closure of several infants with large exomphalos defects including the use of mesh and skin closure alone was published by Olshousen in 1887 but it was in the 20th century that immediate surgical treatment began to be strongly advocated by Tennant in

Glasgow in 1921 and Robert Gross in Boston, USA, who described staged surgical management using skin flaps for closure in 1948. Management with a silo was first described by Sam Schuster in 1967 and this has remained the technique of choice for infants requiring staged surgical closure.

The current alternative, using some form of antiseptic paint, evolved from the use of alcohol described by Ahlfield in 1899 and restated by Cunningham in 1950. Grob and Soave in 1963 popularized conservative management using mercurochrome and published photographic evidence of epithelization of the sac with encouraging results.

EMBRYOLOGY

The term "exomphalos" describes a collection of different abnormalities that have in common the presence of abdominal viscera within an intact sac derived from umbilical cord tissue. Common to all variants is a failure of abdominal wall closure which commences after the end of the third week post-conception when the embryonic disk folds in on itself ventrally in a purse-string like manner. This folding pinches off a length of the yolk sac adjacent to the ventral surface to form the gut tube. The gut tube remains in contact with the yolk sac outside the embryo via a small connection at the umbilical ring, the vitellointestinal duct. Between 6 and 8 weeks post-conception, the gut tube elongates and protrudes outside the embryo before returning to the expanding abdominal cavity to lie in its final position at 10–12 weeks. The muscle layers of the left and right halves of the abdominal wall fuse together above and below the umbilical cord insertion, either through migration of the somatic mesoderm from either side of the developing spine or through the differential growth of areas of the muscular body wall throughout gestation. The umbilical ring remains static in size through gestation due to balanced processes of cell death and cell deposition but reduces in size relative to the fetus. The vitellointestinal duct degenerates so that the cord eventually contains only the umbilical vessels.

PRINCIPLES AND JUSTIFICATION

The relative rarity of exomphalos has meant that historically it has been treated as one condition for the purposes of institutional reporting. However, like other congenital structural anomalies such as ventricular septal defect (VSD) and hydronephrosis, many of the varieties of exomphalos should be thought of as an anatomical end point of underlying genetic anomalies, rather than a "condition" of itself. As such, it differs greatly from gastroschisis, with which it was often, incorrectly, grouped.

Associated genetic abnormalities range from aneuploidy (e.g. Edwards' syndrome, trisomy 18), to smaller genetic aberrations (e.g. Beckwith–Wiedemann syndrome, BWS). Exomphalos itself is a feature of over 70 known malformation syndromes, many with no identifiable genetic cause (**Table 25.1**) and, even in the absence of a syndromic pattern, major structural defects affecting other body systems are common. In a recent UK and Ireland live-born population cohort study using the British Association of Paediatric Surgeons Congenital Anomaly Surveillance System (BAPS-CASS), non-PDA cardiac defects were present in 50% of infants, renal tract anomalies in 13%, other gastrointestinal anomalies in 23%, and limb anomalies in 7%. Intracranial, facial, spinal, and genital anomalies were also detected.

Table 25.1 **Examples of named syndromes or sequences associated with exomphalos**

Named syndrome/sequence	Clinical features occurring in addition to exomphalos
Beckwith–Wiedemann syndrome	Macrosomia, macroglossia, ear abnormalities, hypoglycemia nevus flammeus, and a predisposition to solid embryonal tumors
CHARGE syndrome	**C**oloboma, **h**eart defects, (choanal) **a**tresia, **r**etardation of **g**rowth and development, **e**ar abnormalities and deafness
Pentalogy of Cantrell	Diaphragmatic (midline) hernia, sternal defects, heart and pericardial defects
OEIS complex	**O**mphalocele, (cloacal) **E**xstrophy, **I**mperforate anus, **S**pinal defects
Limb body wall complex/body stalk anomaly	Facial clefts, scoliosis, neural tube defects, and urogenital abnormalities
Donnai–Barrow syndrome	Diaphragmatic hernia, absent corpus callosum, deafness

The live-birth prevalence of exomphalos, which is readily detectable *in utero*, is highly dependent on antenatal counseling practices and rates of termination. In the BAPS-CASS survey, only 34% of antenatally diagnosed infants were born alive, with a resulting prevalence of almost 1 in 10,000 live births.

The size of the actual defect and presence of the extracorporeal liver are usually the factors which determine outcome and type of closure embarked upon. Significant viscero-abdominal disproportion may be related to significant pulmonary hypoplasia and, in some cases, tracheobronchomalacia, either of which can require prolonged ventilatory support. Prematurity may also impact on outcome and can be a major risk factor in those with large defects.

Previous authors have stratified the degree of exomphalos simply according to ”minor” or “major,” typically based on defect size (< or ≥5 cm) and whether the liver is a significant part of the contents, although it should be noted that this has not been agreed universally. Our work with the BAPS-CASS data has shown that the defect diameter (cm)/weight (kg) ratio (DWR) provides a method of describing relative defect size for infants of any birth weight. Thus, we have identified that female infants are more likely to have larger defects than males and that there are a number of infants that are born prematurely with intrauterine growth restriction with relatively large abdominal wall defects.

DWR is related to the presence of the liver in the sac and the ratio that correctly classified whether the liver was in the sac for the highest proportion of children was ≥1.63. Both DWR and extracorporeal liver affected surgeons' ability to close the defect primarily, with the ratio that identified most of those who had staged closure being ≥1.64. Although DWR requires validation in future prospective studies, its use may provide a better standard to report exomphalos characteristics allowing better intra-institutional comparison.

PREOPERATIVE INVESTIGATIONS AND PREPARATION

A careful clinical examination to assess the presence of associated abnormalities is warranted and preoperative echo is recommended for all infants. All infants should have careful blood glucose monitoring after birth due to the association with BWS, though this is much more common with minor rather than major exomphalos. A renal, spinal, and cranial ultrasound should be performed prior to discharge. Parents should be counselled about the value of sending a genetic microarray, which may highlight an underlying cause where one has not already been identified.

Complications

Complications occurring during inpatient stay largely relate to problems associated with comorbidity, most notably prematurity and therefore intraventricular and pulmonary hemorrhage, necrotising enterocolitis, and iatrogenic injury to the sac. Other generic complications may be divided into respiratory, gastrointestinal, and infectious.

RESPIRATORY

Prematurity, pulmonary hypoplasia, and tracheobronchomalacia may lead to a need for prolonged ventilatory support or oxygen therapy. Following discharge, infants may remain highly susceptible to respiratory deterioration in the presence of infection.

GASTROINTESTINAL

Gastroesophageal reflux is a major ongoing problem in some infants related to an abnormal esophageal hiatus, gut rotational anomaly, and increased intra-abdominal pressure following reduction. The need for prolonged tube feeding may lead to oral aversion. Dietetic and gastroenterology input will be necessary where feeding problems arise. Malrotation and non-rotation are known associations, and incidences of volvulus have been described.

INFECTIOUS

Sepsis related to prolonged inpatient stay and indwelling IV catheters may occur. Sac damage during conservative management can be managed with careful suture or biological patch application and tissue glue.

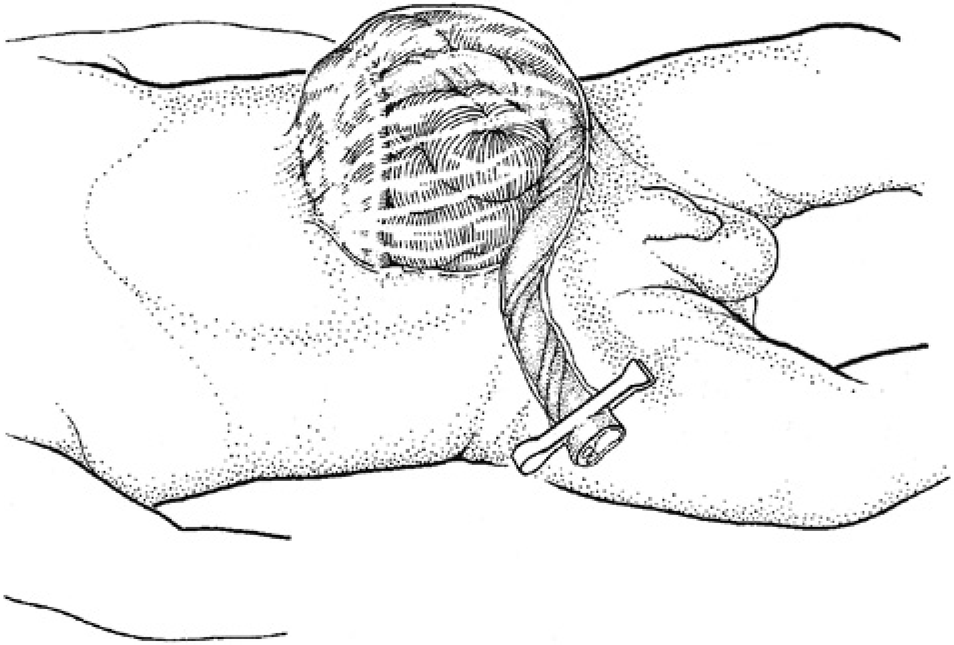
OPERATIONS

Data from BAPS-CASS indicate that virtually all infants with minor (including hernia-into-cord) and many with major exomphalos can be closed surgically.

General anesthesia together with full muscle relaxation is vital. Some surgeons would also try to measure the intra-abdominal pressure (gastric or bladder) objectively, though it is not mandatory. Surgery should not be performed at <24 hours to allow a degree of transitional pulmonary adaptation to occur. Furthermore, it should be regarded as a semi-elective procedure and, if there is a hint of cardiorespiratory instability, it should be deferred for another time. The only absolute indication for urgent surgery is where there has been major sac disruption. Prophylactic antibiotics are usual, particularly if prosthetic material is to be used.

Minor exomphalos

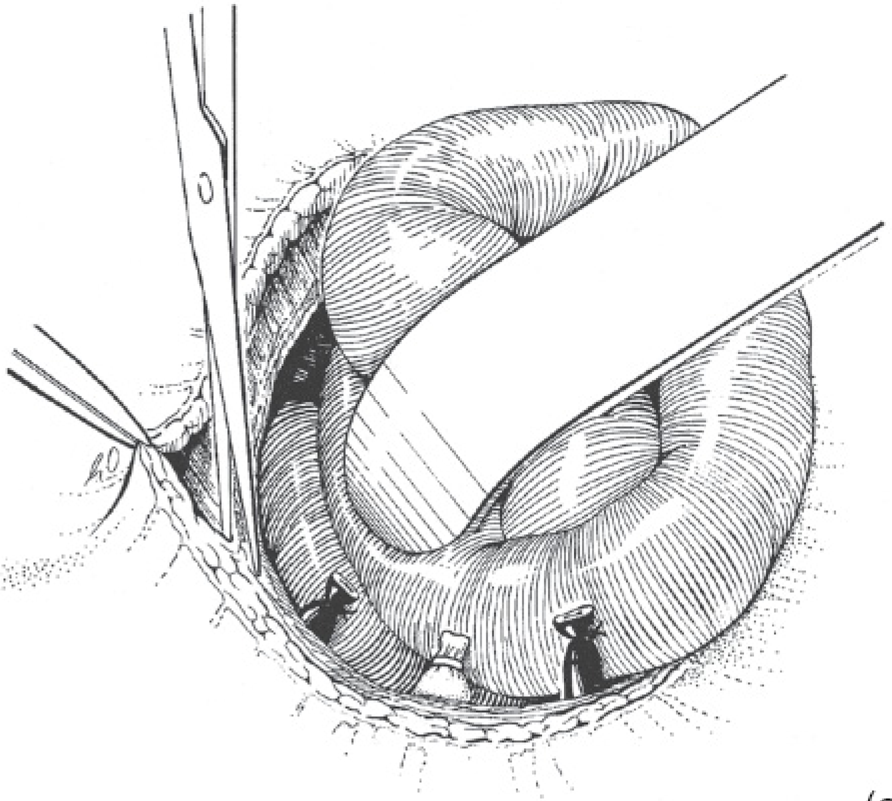
The sac is entered through a convenient window and the umbilical vein and two arteries should be detached from the inside (**Figure 25.1**). The contents should simply be the intestines, or at the most a tongue-like projection of liver. A Meckel's diverticulum representing the apex of the midgut loop may be stuck and may also need separating.



25.1

The main decision to be made is the rotation status of the midgut loop. If this is not obvious from the contents, then the fascial defect can be widened in any dimension to allow the duodenojejunal (DJ) junction and ileocecal (IC) valve to be located. Normal rotation and complete non-rotation require nothing more than documentation; however, correction of malrotation by widening of the mesentery and Ladd's procedure may be warranted. Some surgeons recommend a preoperative contrast meal and follow-through to at least help identify the position of DJ flexure.

Having returned all intestines to lie comfortably with the cavity, the layers of the fascia must be separated and closed accordingly. Enough sac and skin is excised to leave enough to close leaving a decent neoumbilical appearance. A subcutaneous purse-string achieves central position, and inversion of small skin flaps leaves a suitably puckered appearance (**Figure 25.2a and b**).



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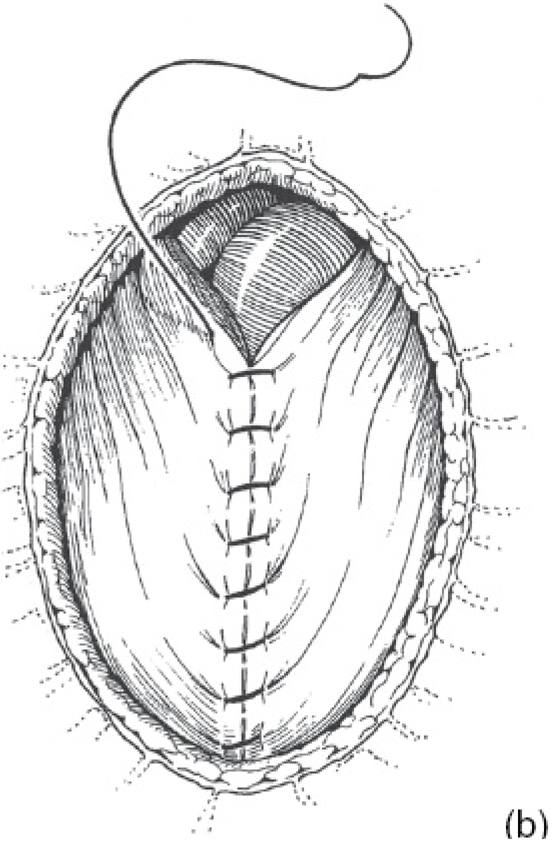
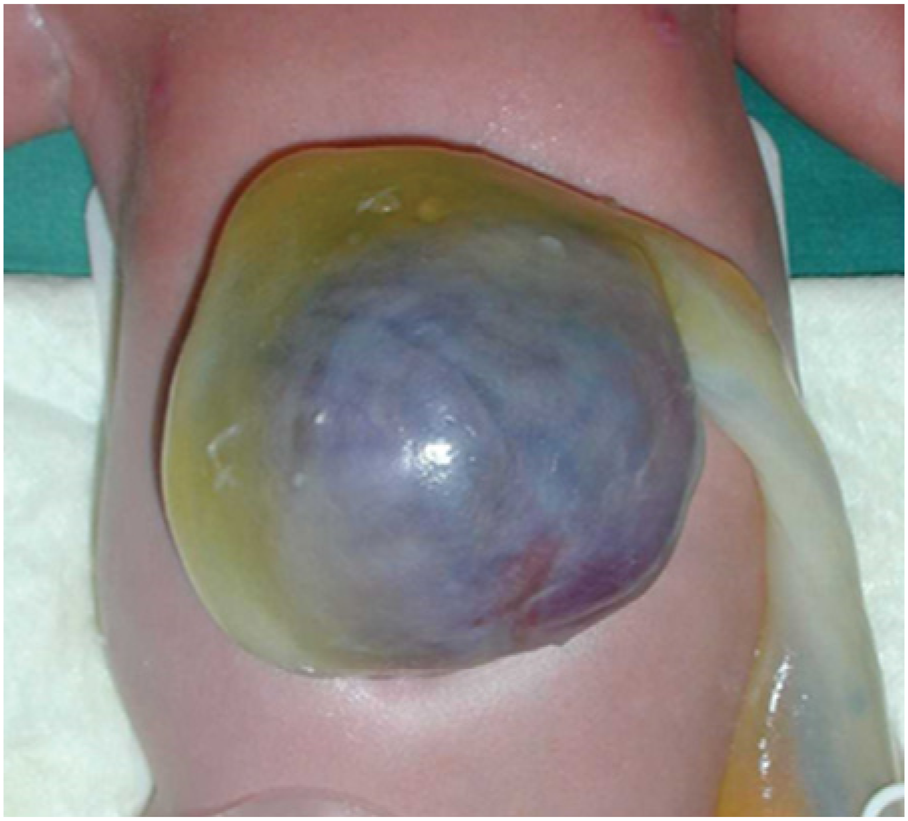


(b)

25.2a,b

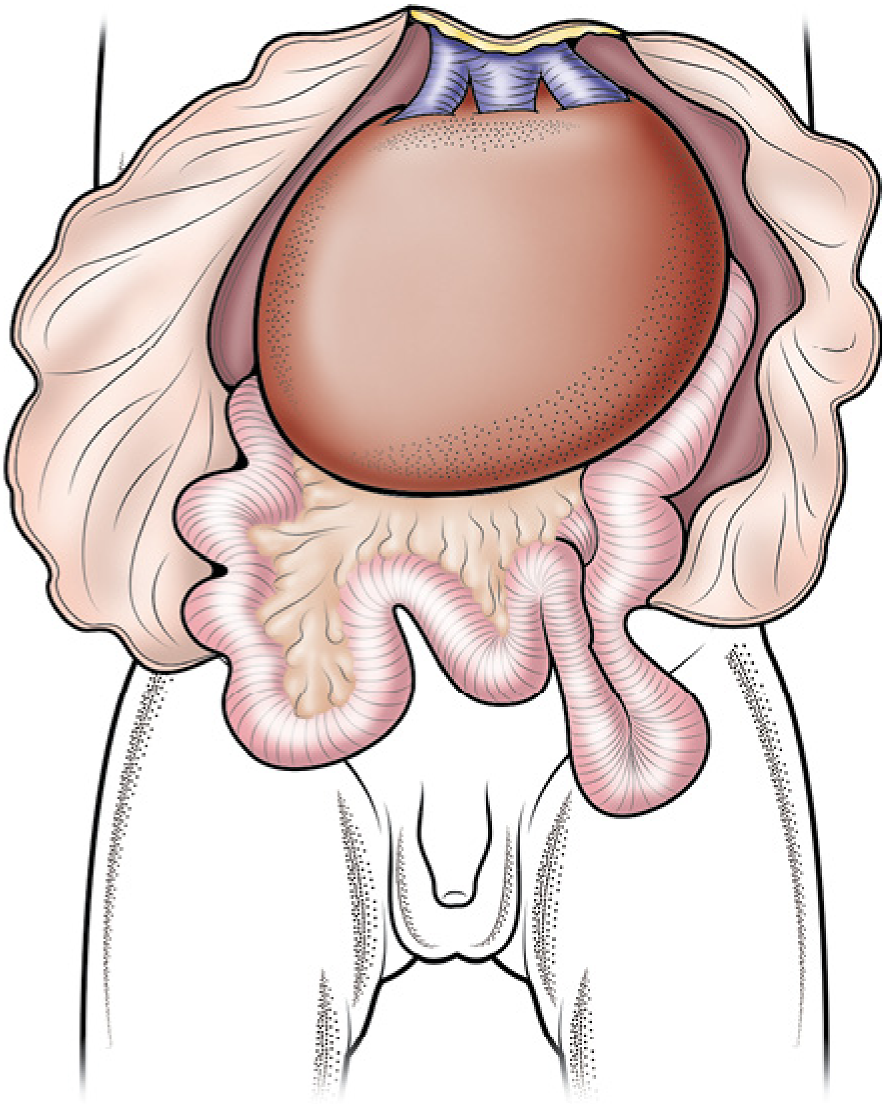
Major exomphalos

Skin preservation is crucial in major exomphalos. Leave any excision to the very end. Simply open the sac distally, entering the peritoneal cavity (**Figure 25.3**). Detach the umbilical vein, and two umbilical arteries from the inside. Confirm the non-rotational status of the intestines (usual finding) and divide any congenital adhesions.



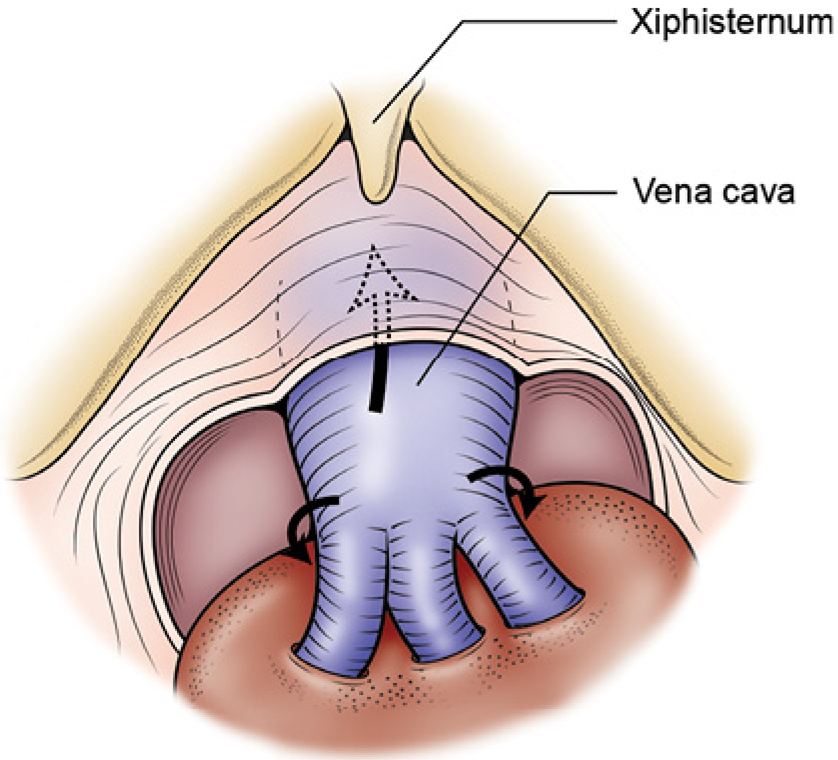
25.3

The focus is on mobilization of the liver (**Figure 25.4**). The sac can be dissected free of the capsule using bipolar diathermy but, if this is causing too much bleeding, then the situation should be accepted and an island of sac and capsule left intact.



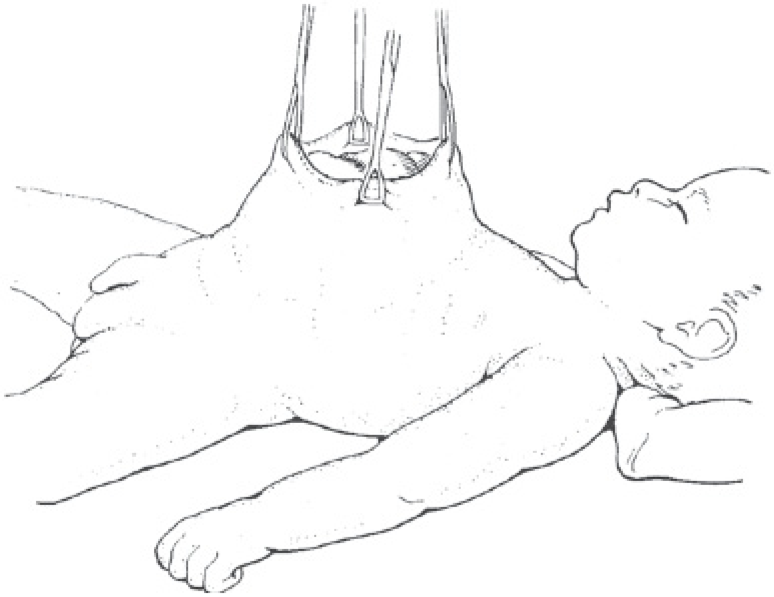
25.4

Whole-liver eventration will inevitably cause the posteriorly placed inferior vena cava to have a “humpback” course and awareness of this is crucial. The hepatic venous outflow is also extended superiorly and this needs careful mobilization from the region of the xiphisternum (**Figure 25.5**). The dissection is started on either side of the hepatic veins, dividing any suspensory ligaments retaining it to the anterior fascial ring.



25.5

This should also allow the liver to drop back into the abdominal cavity occupying the right sub-diaphragmatic space. Counter-traction with stay sutures or Babcock forceps placed around the margins of the sac or fascia helps ease this key stage (**Figure 25.6**). Sometimes the size of the actual defect is the limiting factor. If so, then lateral full-thickness division may overcome any difficult of actual organ reduction. Again, be aware of the fragility of the liver capsule and parenchyma with the propensity to cause liver bleeding. Enveloping the exposed liver tissue with Surgicel^{TM} hemostatic weave may be useful.



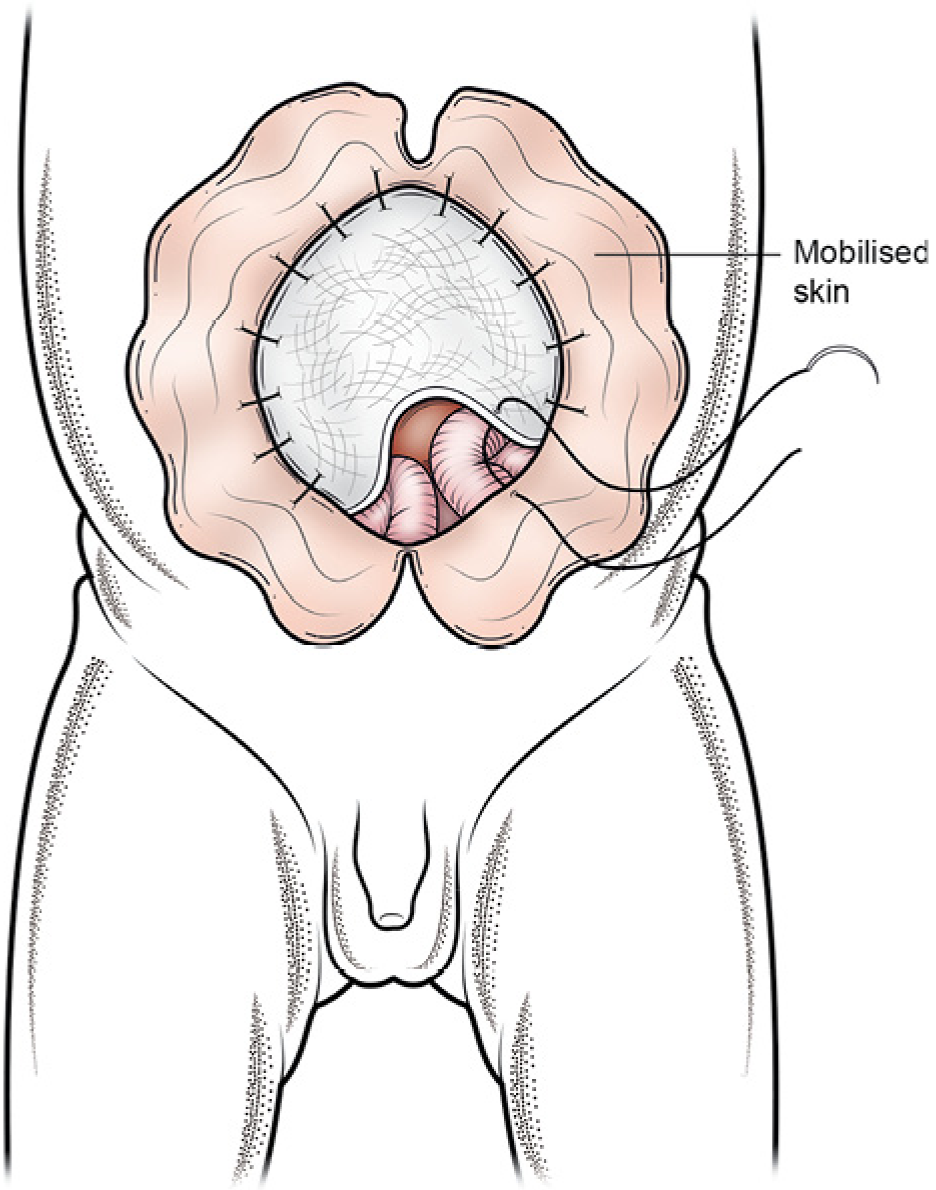
25.6

Once the liver is within the cavity the position of the much more complaint intestines can be considered. Be aware of the nature of the axis of the liver (i.e. hepatic vein, liver, portal vein/artery), maintaining its linearity without kinking or compression. It should be possible to trace this axis through the superior mesenteric vein to the edge of the small bowel in those with a non-rotated midgut.

Try to maximize the abdominal wall domain by finger-stretching its inside. Reduce the intestines, again avoiding untoward rotation or volvulus. If, in the end it all looks good, accept the new situation and wait. Ensure ventilation remains easy and blood gases appropriate.

The next decision is to whether the fascia can now be closed primarily. If thought safe to do so, separate the skin from the fascia and close the latter in the direction (vertical or horizontal) that seems most appropriate. At the end, again check the situation with the anesthetist.

If this is difficult, not possible, or the closure appears too tight, go back a stage and use a prosthetic patch (e.g. Permacol®, Gore-Tex®) sewn in and overlapping into the cavity with non-absorbable sutures (**Figure 25.7**). The key to successful patch incorporation is good skin cover.

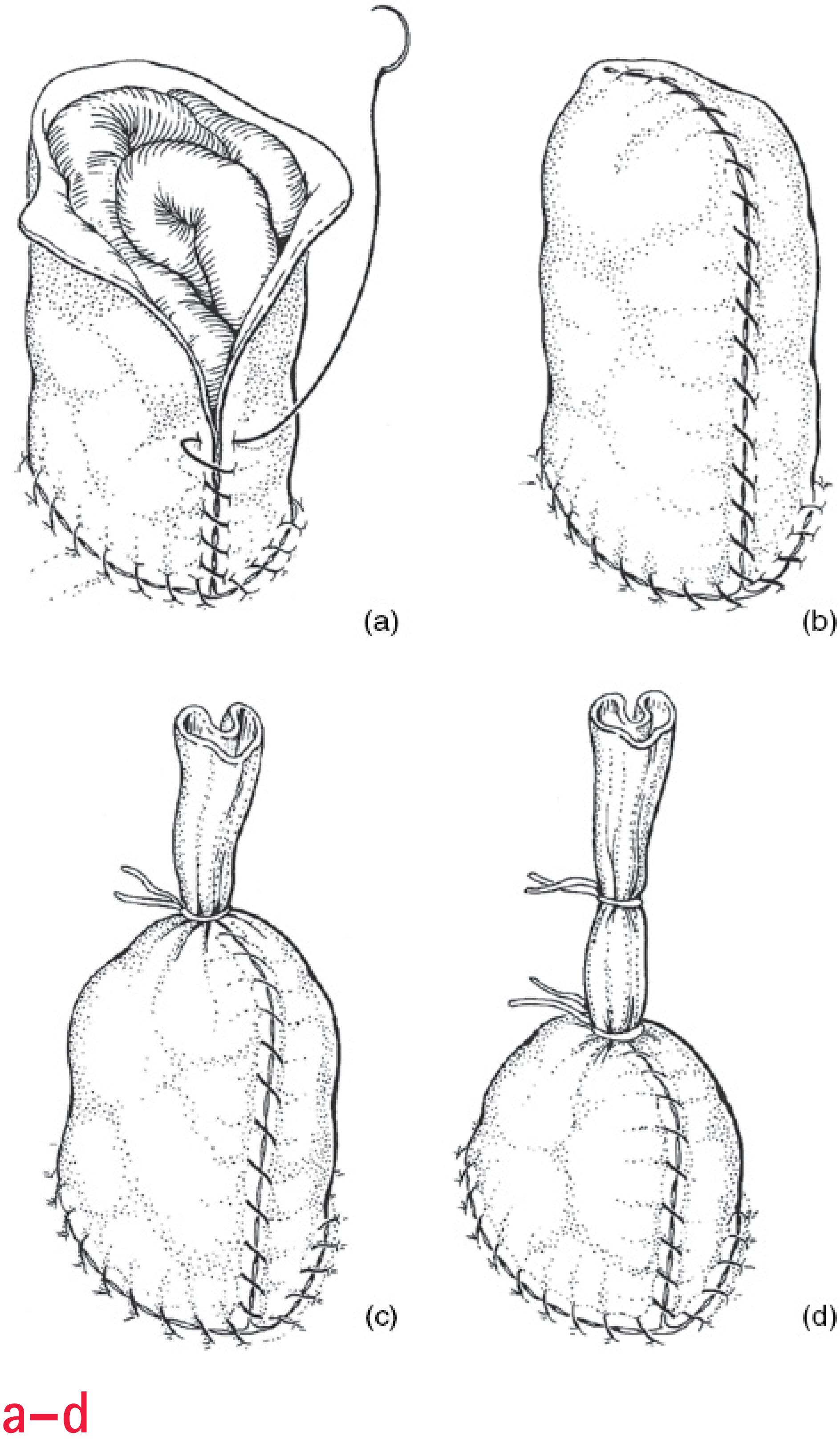


25.7

Silo closure

For those where primary closure is not possible, with or without a patch, a period of time in a silo is appropriate. It has been our practice to perform the liver mobilization described above during the first stage to facilitate visceral mobility within the silo later.

Again, there are many options, including single-layer, double-layer, custom, and preformed. Whatever is chosen, it is important to end up with a robust system which will allow serial tightening or reduction of the silo to push visceral content back into the abdominal domain (**Figure 25.8**). Typically, a flat silo closure should be achieved over a week in a ventilated infant. During the latter stages, try to pull the opposite sides of the fascial edge toward each other, making the definitive surgery easier.



25.8a–d

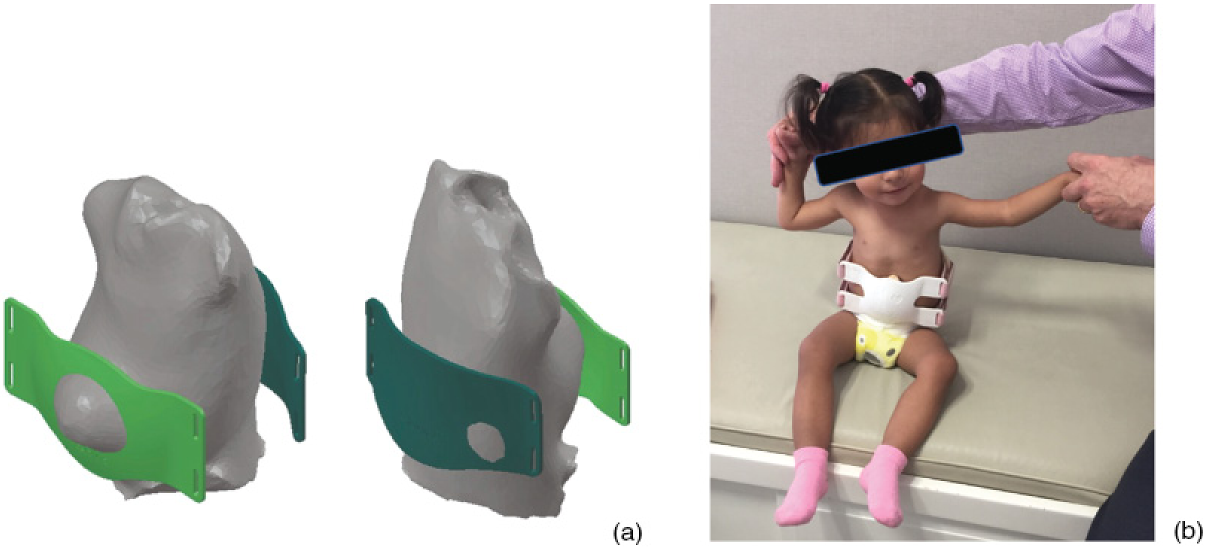
Conservative management and secondary closure

For those infants treated conservatively with sac preservation for one reason or another, it is important to then plan for the definitive repair and embark on a realistic timescale with the parents (**Table 25.2**) (**Figure 25.9a and b**).

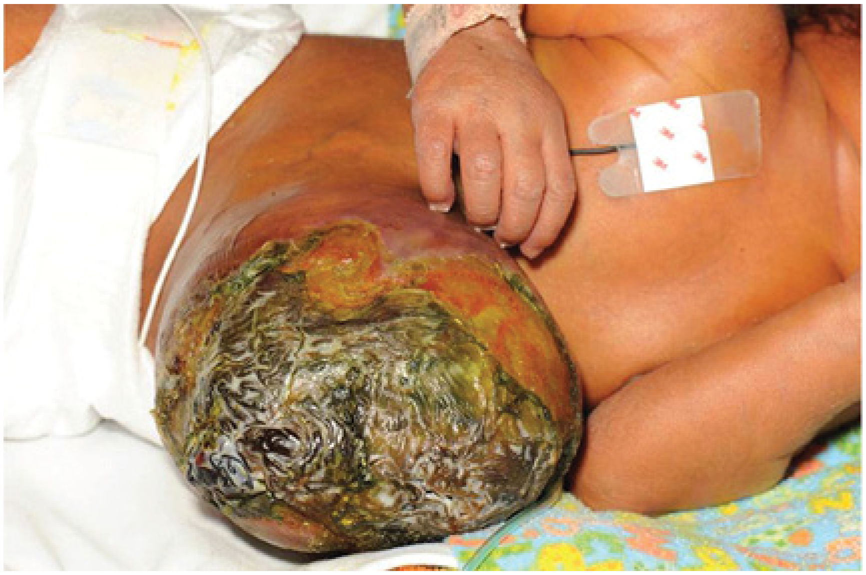
Table 25.2 **Conservative management of exomphalos**

This technique may be employed in infants with significant viscero-abdominal disproportion or comorbidity precluding initial surgical management.

- Use of topical antiseptic applied to the sac is popular. Silver sulfadiazine cream 1% was the most commonly used therapy in the BAPS-CASS study with no apparent ill effects. Alternative treatments include manuka honey, Betadine™, etc. "Dry management" is also feasible.
- Dressings and bandages with padding to support the sac are commonly used.
- Regular bathing of the infant is encouraged when the epithelialized sac has become robust (Figure 25.10).
- The infant should be allowed to feed as early as possible, ideally from the breast.
- Prophylactic antibiotic therapy is not warranted but careful attention should be paid to signs of sac or skin infection.
- Discharge from hospital can occur when the infant is medically stable and before full epithelialization of the sac.
- Some surgeons favor compression in the form of elasticated abdominal supports prior to definitive closure.
- Timing of definitive closure depends on the infant's general condition and development needs and the wishes of the family.



25.9 “Argentine Sandwich”. Courtesy of Dr Luzía Toselli, Buenos Aires.



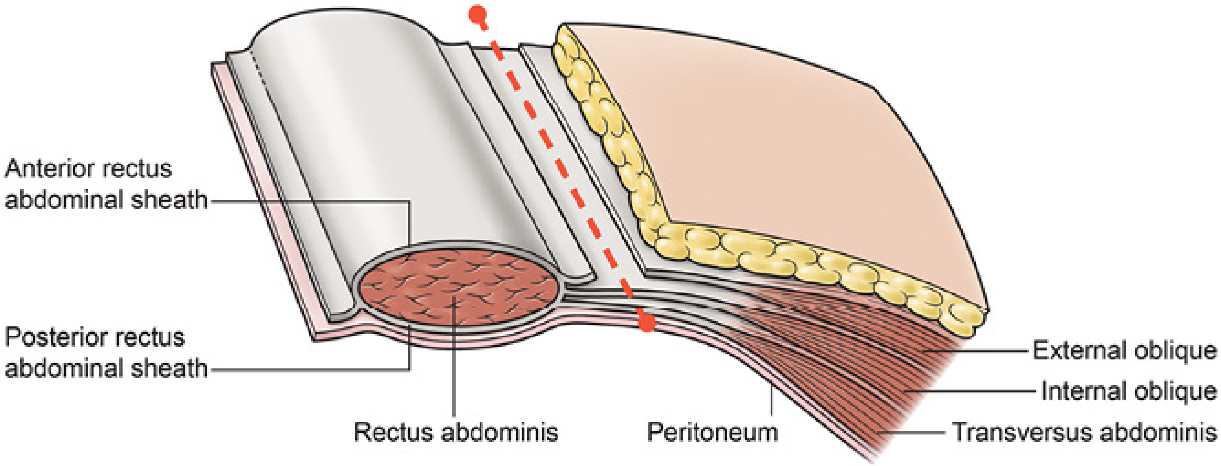
25.10 Conservative treatment: appearance of exposed sac at 6 weeks.

This may take only a year but in some patients extends to 4–5 years or even longer. During this hiatus, the protuberant abdomen needs protection and support. This should be customized to the patient. There are intriguing devices available involving 3D printing and precise body measurements. In principle and practice, these should also apply external pressure, again facilitating later definitive repair.

Although in the long term most secondary defects can be relatively straightforward to close surgically, again with or without prosthetic patches, it should be remembered that the child still has to tolerate the inevitable increased intra-abdominal pressure, diaphragmatic splinting, etc. Arrangements for postoperative ventilation and appropriate monitoring need to be in place to avoid compartment syndrome.

Component separation technique

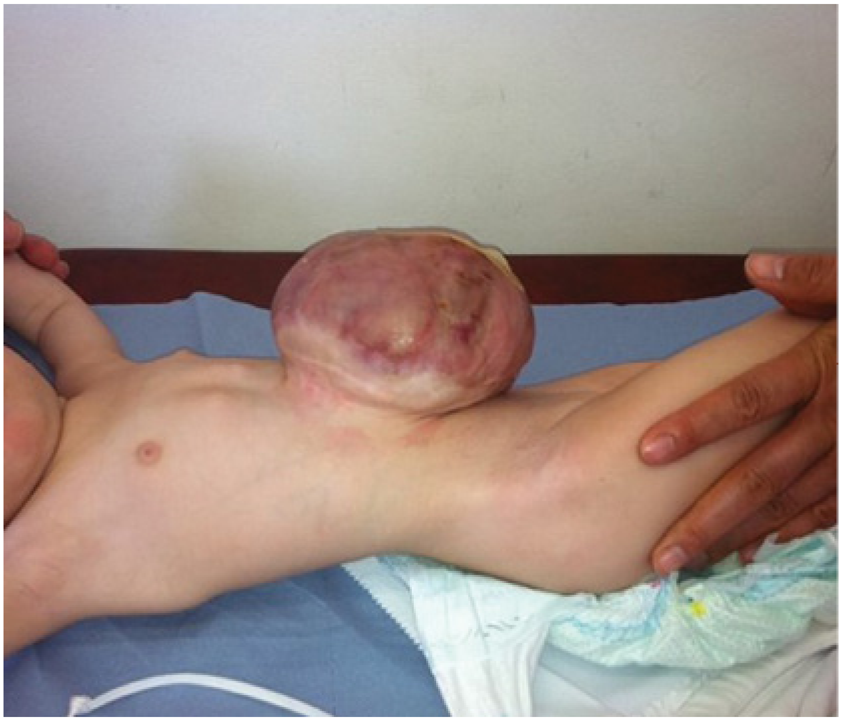
The component separation technique is suitable for the delayed reconstruction of the abdominal wall and may avoid prosthetic patch use entirely. It involves undermining of the skin until the junction of the anterior rectus sheath and external oblique and then longitudinal incision of the external oblique anastomosis lateral to the rectus. This then allows the underlying muscles of the internal oblique and transversus to slide medially, facilitating midline closure or reduction of wall tension (**Figure 25.11**).


Anterior rectus abdominal sheath
Posterior rectus abdominal sheath
Rectus abdominis
Peritoneum
External oblique
Internal oblique
Transversus abdominis

25.11 Component separation technique.

Minimal abdominal domain

Some infants have an extreme form of limitation of abdominal domain. Sometimes the defect actually is small but the eviscerated organs are large. This results in a more "mushroom-like" external appearance (**Figure 25.12**), precluding any kind of preoperative reduction. For these children, skin cover may be the limiting factor to a successful abdominal wall closure. Consideration should therefore be given to insertion of subcutaneous tissue expanders prior to an attempt at closure.



25.12 Delayed treatment awaiting definitive repair.

OUTCOME

The outlook for the majority of infants born with exomphalos is excellent.

In the BAPS-CASS study of 162 infants, 59% underwent successful primary defect closure. Where staged closure was required (*n* = 37), surgeons favored conservative management, with 23 (62%) using this treatment when they had a choice, i.e. with infants who were fit for general anesthesia and had an intact sac.

Survival to 28 days from surgery or decision for non-operative management was achieved in 93% of infants. Three of the seven non-survivors of this early period received comfort care due to significant associated comorbidity. By 1 year, 87% (106/122) of the cohort were known to be still alive. Determinants of survival included prematurity, birth-weight intrauterine growth restriction, the presence of other anomalies, sac rupture, and DWR.

Median (interquartile range) length of initial hospital stay for all infants in the cohort was 13 (8–40) days, and was 10 (5–13) days, 31 (18–71) days, and 78 (40–153) days for those undergoing primary closure, conservative management, and silo reduction respectively. Fifteen of 106 infants who survived to 1 year (14%) were receiving respiratory support at 28 days after birth. Twelve of these 15 infants were born preterm, and 3 were still receiving support 1 year after birth.

Assessment of outcomes of infants born with exomphalos, particularly in relation to optimal management choice for those requiring staged closure, should include the outcomes of the later ventral hernia repair. Very few accounts of longer-term outcomes of exomphalos management exist, including those reported by patients and parents. Those that do have highlighted cosmesis and abdominal pain as possible long-term concerns.

FURTHER READING

BAPS-CASS website: www.npeu.ox.ac.uk/baps-cass (accessed 10 April 2020)

Christison-Lagay ER, Kelleher CM, Langer JC. Neonatal abdominal wall defects. *Semin Fetal Neonatal Med* 2011; 16(3): 164–72.

Lakasing L, Cicero S, Davenport M, et al. Current outcome of antenatally diagnosed exomphalos: An 11 year review. *J Pediatr Surg* 2006; 41: 1403–6.

Rijhwani A, Davenport M, Dawrant M, et al. Definitive surgical management of antenatally diagnosed exomphalos. *J Pediatr Surg* 2005; 40: 516–22.

van Eijck FC, van Vlimmeren LA, Wijnen RM, et al. Functional, motor developmental, and long-term outcome after the component separation technique in children with giant omphalocele: A case control study. *J Pediatr Surg* 2013; 48: 525–32.

26

Gastroschisis

MARSHALL Z. SCHWARTZ AND SHAHEEN J. TIMMAPURI

HISTORY

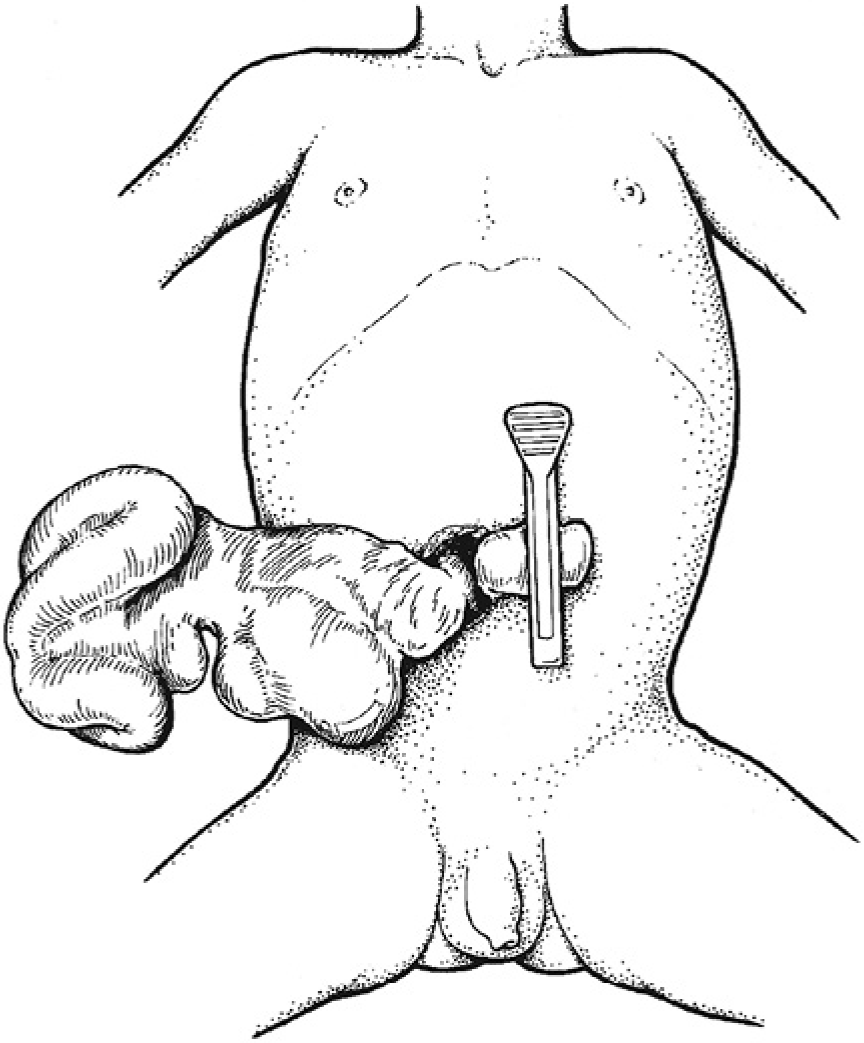
The first successful surgical repair of gastroschisis was performed by Watkins in 1943. Although there were improvements in the perioperative management and surgical procedures for gastroschisis, the mortality remained significant and was reported to be as high as 90%. Two major advances occurred in the late 1960s that led to a dramatic improvement in the survival of infants with gastroschisis. In 1967, Schuster et al. described a technique of staged reduction of the herniated bowel and abdominal closure for patients in whom primary closure was not possible. The second major advance was the evolution of IV nutrition, which, remarkably, allowed for growth and development during the prolonged period that these infants would not tolerate enteral feeding.

Over the ensuing three decades and to this day, the outcome for infants with gastroschisis has dramatically improved. The survival rate is now greater than 90%.

PRINCIPLES AND JUSTIFICATION

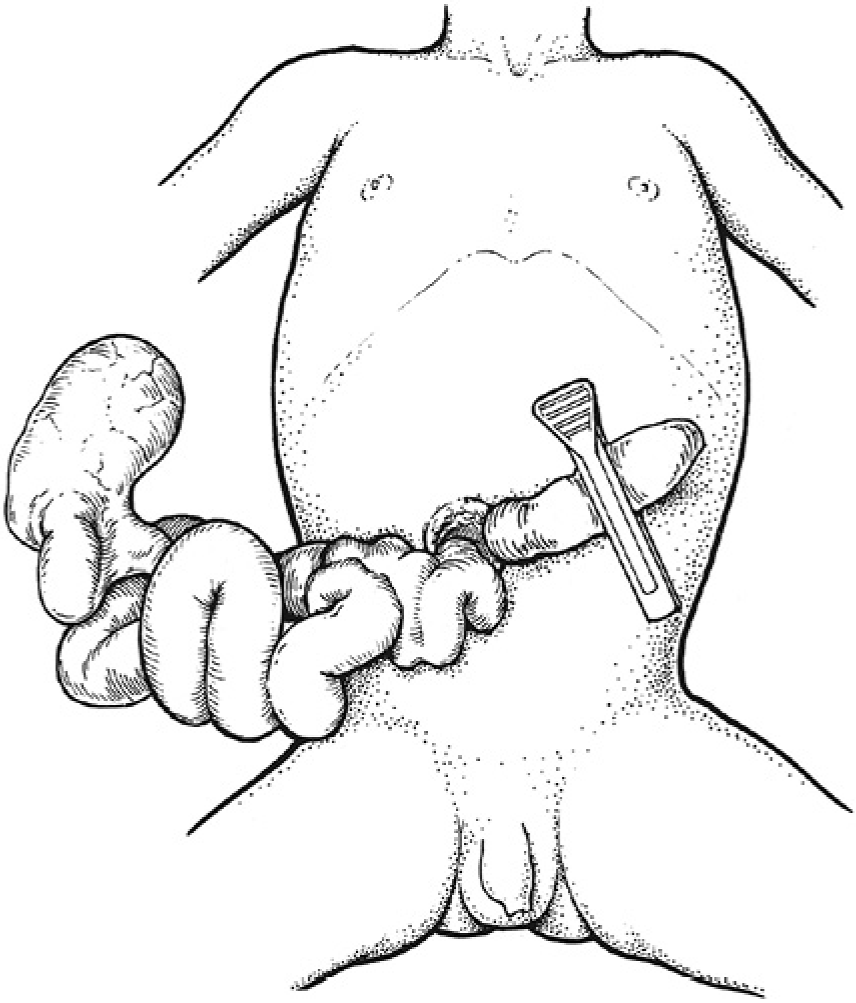
Gastroschisis is an anterior abdominal wall defect that occurs *in utero*, through which there is herniation of intra-abdominal viscera into the amniotic sac. It is thought to result from a defect at the site of involution of the second (right) umbilical vein. This anomaly is accompanied by non-rotation of the bowel and an increased incidence of intestinal abnormalities, including atresia, perforation, and infarction, resulting from midgut volvulus or vascular thrombosis. The incidence of gastroschisis is approximately 1 in 4000–6000 live births.

Most infants with gastroschisis are born prematurely (35–37 weeks' gestation), weighing 2000–2500 g. The defect is almost always to the right of the umbilicus and generally measures 2–3 cm in diameter. All or a portion of the midgut is usually herniated through the defect (**Figure 26.1**). In addition, the stomach, urinary bladder, and, in females, the Fallopian tubes and ovaries may also be extracelomic.



26.1

The intestine is foreshortened, edematous, and generally has a fibrin coating. Atresia of the small and large intestine occurs more often (approximately 14%) than in patients with omphalocele (1%). The most striking difference in appearance between omphalocele and gastroschisis is the absence of a sac or membrane covering the herniated contents in gastroschisis (**Figure 26.2**). Continuous contact of the herniated contents with amniotic fluid in the third trimester has been proposed as the reason for the thickened, foreshortened, and edematous bowel. There is also increasing evidence that the fascial defect begins to decrease in diameter near the end of the third trimester, which could lead to venous congestion of the midgut and contribute to the abnormal appearance of the bowel at birth. Therefore, serial fetal ultrasonography in the third trimester is warranted to follow the appearance of the bowel. Bowel dilatation and echogenic bowel have been described as possible predictors of postnatal morbidity. Progressive worsening of these findings could lead to future complications and may be an indication for early delivery. Routine Cesarean section is not recommended for infants with gastroschisis.

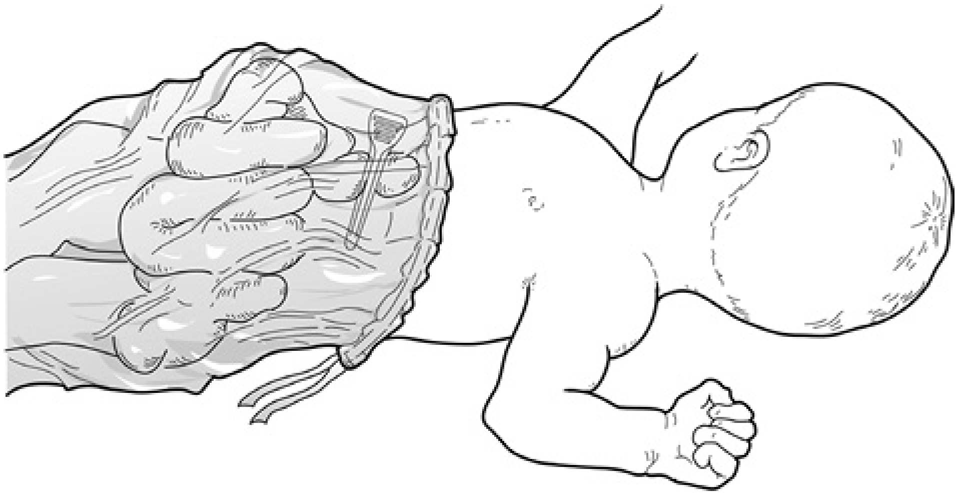


26.2

PREOPERATIVE ASSESSMENT AND PREPARATION

Gastroschisis requires prompt surgical intervention. Delays in surgical management should be incurred only as a result of transport of the infant to a pediatric surgical center or the need for prolonged preoperative stabilization. The evaluation of other life-threatening anomalies, which are rare, may delay surgery. Appropriate preoperative preparation is essential to ensure a good outcome.

Maintenance of the infant's temperature within the normal range is critical because heat loss from the exposed herniated contents can be significant. There are numerous methods to minimize this heat loss depending on what supplies are available in the delivery room or intensive care nursery. Damp, warm gauze sponges or a damp, warm roll of gauze is wrapped around the intestine. Next, a plastic bag containing the baby from the feet to the chest may be used to maintain normal body temperature and diminish evaporative fluid loss (**Figure 26.3**). Alternatively, dry rolls of gauze may be wrapped around the patient's abdomen and the damp gauze-covered bowel to create an appropriate environment to retain heat and minimize evaporative fluid loss. It is imperative to stabilize the bowel and therefore diminish the risk of compromising its blood supply at the fascial ring. This can be accomplished by placing the infant on his/her side. The infant should be in a warming isolette or under an overhead radiant warmer to help maintain normothermia.



26.3

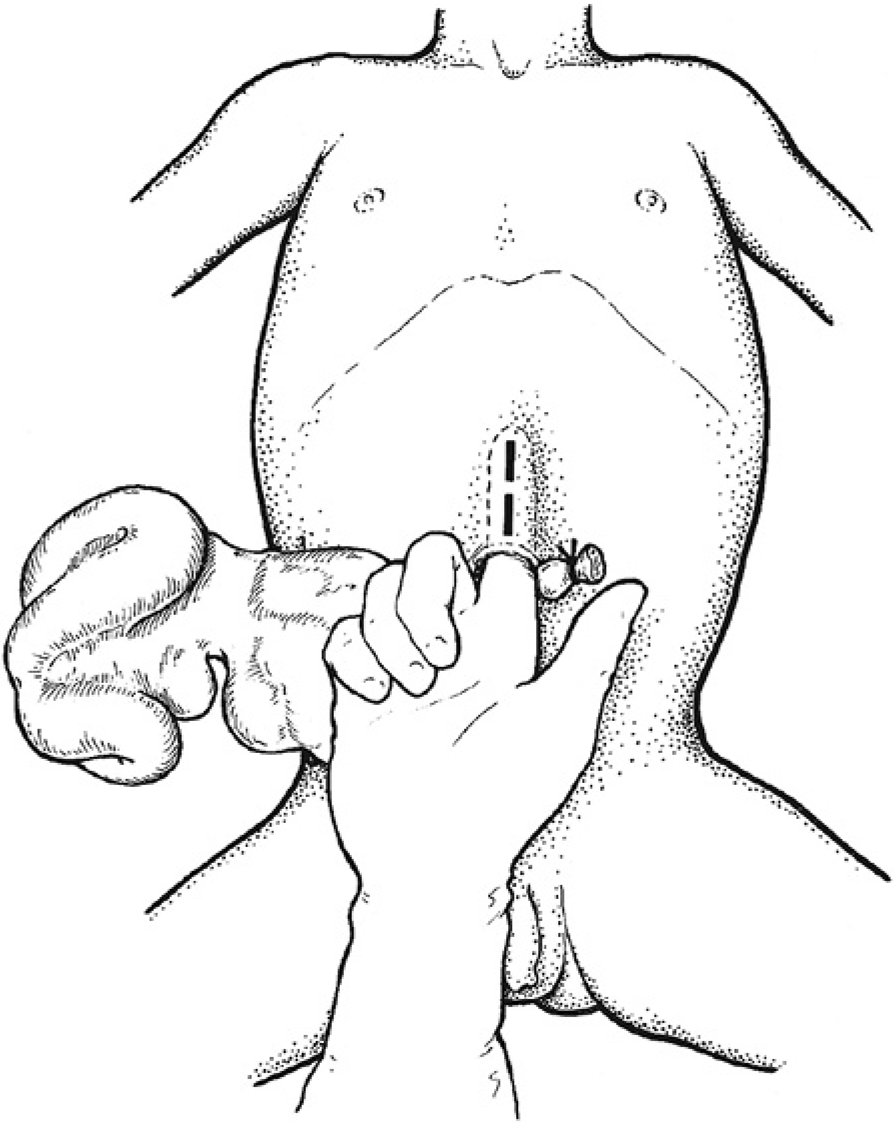
Most infants with gastroschisis are dehydrated at birth and require at least 125% of normal maintenance fluids to regain normovolemia. Eventually, almost all infants with gastroschisis will require central venous access. This can be accomplished by placement of a tunneled-cuffed catheter or a peripherally inserted central catheter (PICC). However, attempts are made to delay the placement of a tunneled central venous catheter (CVC) until 2–3 days after surgery to decrease the risk of catheter contamination by bacterial translocation or other sources. It is appropriate to give broad-spectrum antibiotics during the perioperative period because of exposure of the bowel and peritoneal cavity to bacterial contamination at the time of birth. A nasogastric tube placed at the time of birth is necessary for gastrointestinal decompression because of the bowel inflammation and resulting ileus.

Thus, before surgery, the infant with gastroschisis should be normothermic, hemodynamically stable, have good urine output, and have normal serum electrolytes following adequate fluid resuscitation.

Anesthesia

General anesthesia is required for appropriate operative management of gastroschisis. The choice of anesthetic agents should be made by the anesthesiologist, but two points should be emphasized: First, muscle paralysis is useful in optimizing the chances for complete reduction of the herniated bowel and primary abdominal wall closure, and second, nitrous oxide should not be used as it diffuses into the lumen of the bowel causing distension and compromising the likely success of complete reduction of the herniated bowel into the peritoneal cavity and primary abdominal wall repair.

It is sometimes necessary to extend the abdominal wall defect to facilitate reduction of the herniated bowel. This is generally done by extending the defect superiorly in the midline by 1–3 cm (**Figure 26.4**). Extending the incision caudally is not recommended because the urinary bladder is in close proximity to the inferior aspect of the abdominal wall defect. The length of this incision depends on the size of the original defect and the bulkiness of the herniated bowel.



26.4

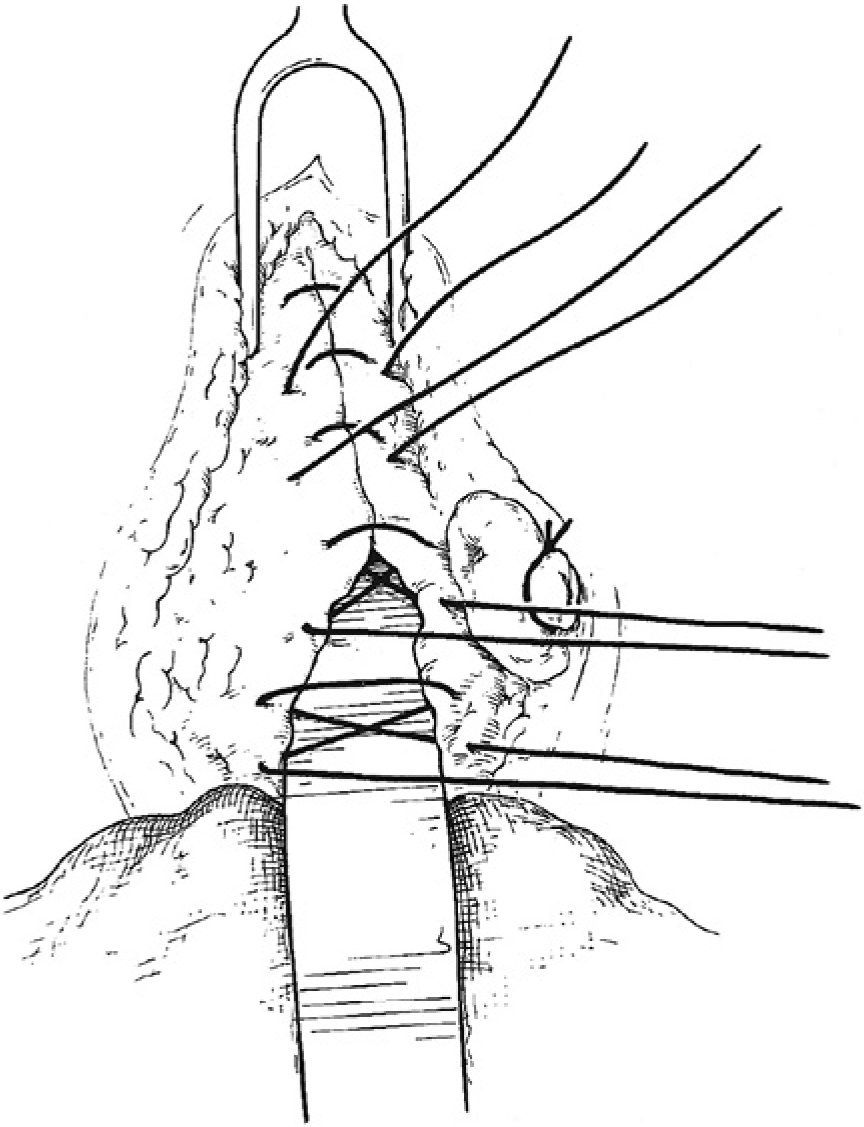
OPERATION

The operation should be performed under conditions that maintain normothermia. Several methods exist to accomplish this goal. An overhead radiant warmer, warming lights, or a warming blanket placed under the infant should be used to maintain the infant's temperature in the normal range during the procedure. Raising the temperature in the operating room may also be necessary. After the induction of general anesthesia, the dressing previously placed over the herniated contents should be removed.

The bowel should be handled with sterile gloves. The umbilical cord, which has usually been left long, should be clamped 2–3 cm above the abdominal wall and the excess cord then removed. Holding the bowel and clamp on the umbilical cord in one hand, the bowel should be prepared using gauze sponges soaked in a 50 : 50 mixture of povidone iodine solution and saline. The antiseptic solution must be warm to the touch to minimize heat loss. After gently washing the bowel and the anterior and lateral abdominal wall, drapes are appropriately placed, and the herniated contents are laid on the drapes. The surgeon should then scrub and put on gown and gloves. The umbilical cord can now be tied below the previously placed clamp with a heavy suture or umbilical tape, allowing removal of the clamp. Next, the herniated intestine should be carefully inspected for areas of perforation or sites of atresia, although no effort should be made to dissect matted loops of intestine.

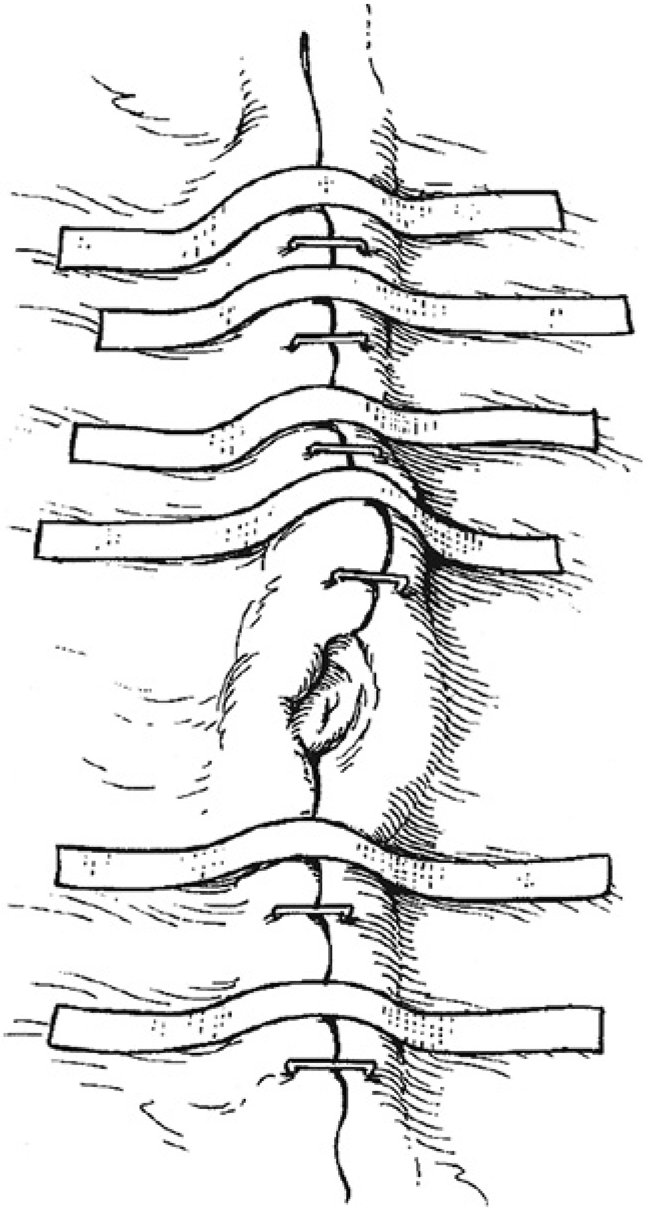
The herniated intestine is reduced as much as possible, distributing the bowel to all quadrants of the peritoneal cavity. Two techniques have been described to facilitate complete bowel reduction and abdominal wall closure: Stretching of the anterior abdominal wall, and "milking" the intestinal contents into the stomach where they can be aspirated through the nasogastric tube. Some surgeons also find milking out the colonic contents to be an effective maneuver for decompressing and reducing the bowel. Although gentle stretching of the anterior abdominal wall can be useful, the authors are opposed to vigorous stretching. This maneuver can lead to rectus muscle hemorrhage and abdominal wall edema, producing a non-compliant, firm anterior abdominal wall, resulting in ventilation difficulties and wound-related problems postoperatively. Also, the authors do not advocate manipulating the intestine to "milk" the intestinal contents into the stomach, believing that this can cause further damage to the bowel wall with serosal tears and resulting in increased bowel wall thickening and additional delay in bowel recovery.

If reduction of the herniated intestine is successful, the abdominal wall is assessed for primary closure. If it can be closed without undue tension, 3/0 absorbable, monofilament sutures are used. These sutures are placed in a figure-of-eight, as this results in fewer knots (**Figure 26.5**). When all the sutures have been placed, they are tied in sequence with a thin, malleable retractor initially underneath the fascia to prevent a loop of intestine from becoming entrapped under the sutures. The umbilical stalk is retained to create a more natural umbilical appearance when the wound is fully closed.



26.5

When the fascia has been closed, the skin edges are approximated using interrupted absorbable sutures or a few skin staples and sufficient sterile skin closure strips, allowing distribution of skin tension over a wider surface area of the anterior abdominal wall and thus reducing the likelihood of skin disruption (**Figure 26.6**).



26.6

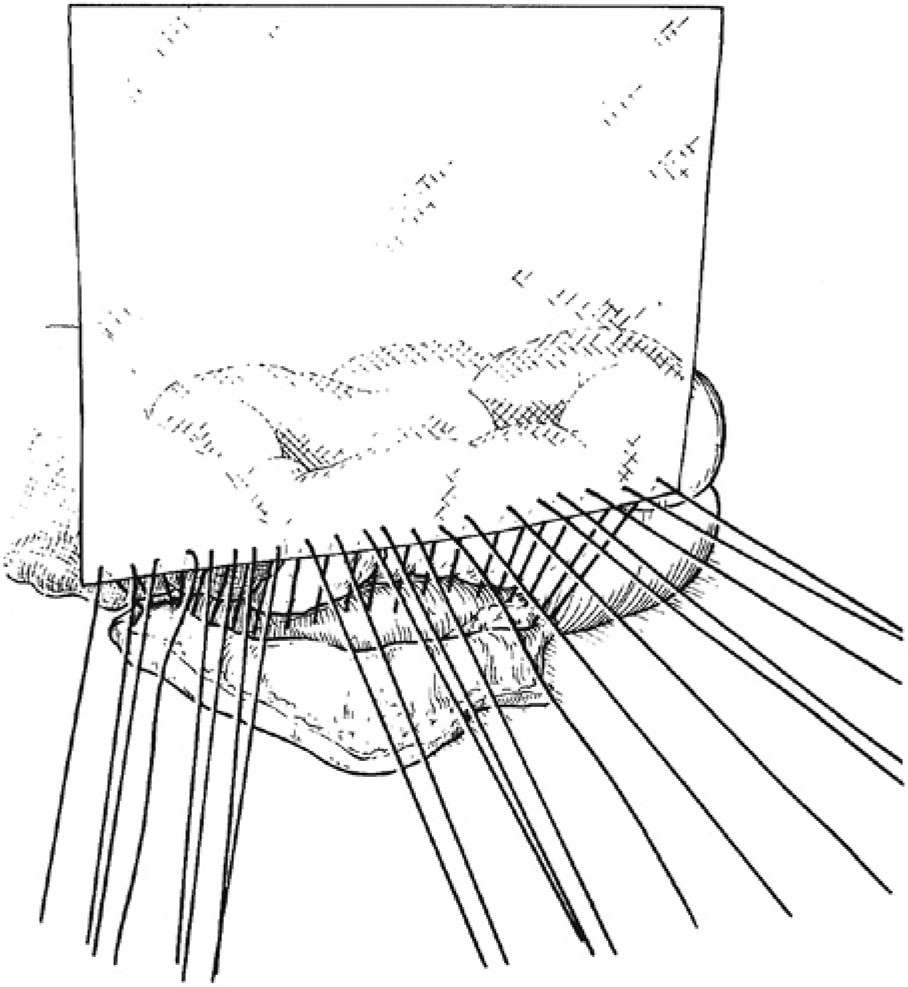
Figure 26.7 shows the appearance 1 week after primary closure. Note that the umbilicus was retained. **Figure 26.8** shows the appearance 3 weeks after primary closure.

26.7

26.8

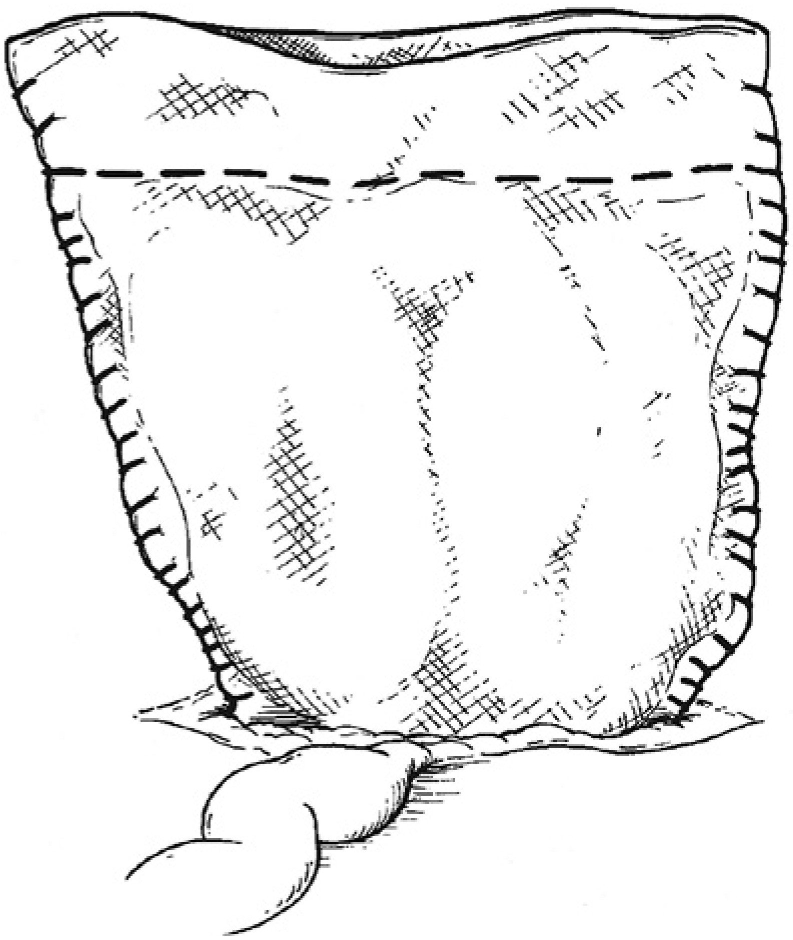
About 60–70% of infants with gastroschisis can be operatively treated in this way without creating undue intra-abdominal pressure or tension in the abdominal wall closure. It is best to avoid high intra-abdominal pressure and excessive suture line tension. This can result in abdominal compartment syndrome, possibly leading to intestinal necrosis, renal hypoperfusion, and difficulty in ventilation, as well as wound disruption. Intragastric and bladder pressure monitoring has been used by some pediatric surgeons to measure intra-abdominal pressure and used as a guide to avoid excessive intra-abdominal pressure during primary or staged closure of gastroschisis. The goal of therapy is to maintain intra-abdominal pressure below 20 mmHg. This is based on prior studies showing that higher pressures compromise intra-abdominal organ perfusion.

For patients in whom complete reduction of the herniated bowel and abdominal wall closure are not possible or appropriate, the staged reduction technique described by Schuster in 1967 has proved to be very useful. Reinforced Silastic sheeting (0.8–1.0 mm thick) is sutured to the fascial edges (**Figure 26.9**). This is accomplished with interrupted 3/0 silk mattress sutures. It is generally necessary to enlarge the fascial defect prior to suturing the Silastic sheet. However, extending the fascial opening too far inferiorly should be avoided as bladder injury may occur.

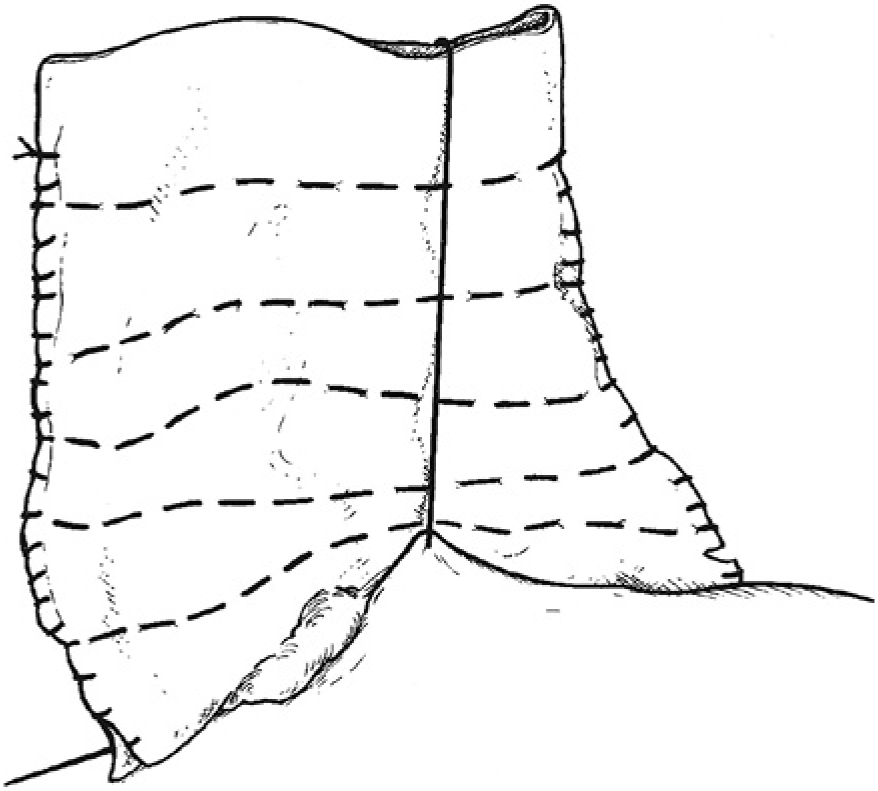


26.9

The cephalad and caudal vertical edges of the silo are constructed with running 3/0 monofilament sutures. Before closing the top of the silo, as much of the bowel as possible is reduced into the peritoneal cavity by manual compression within the silo while avoiding excessive intra-abdominal pressure. The top of the silo is oversewn with a 3/0 monofilament suture placed in a running horizontal mattress fashion (**Figure 26.10**). A suture is also placed through the skin edge laterally midway between the cephalad and caudal limits of the abdominal wall defect and looped over the silastic silo and sutured through the skin on the other side (**Figure 26.11**) to minimize skin retraction.



26.10

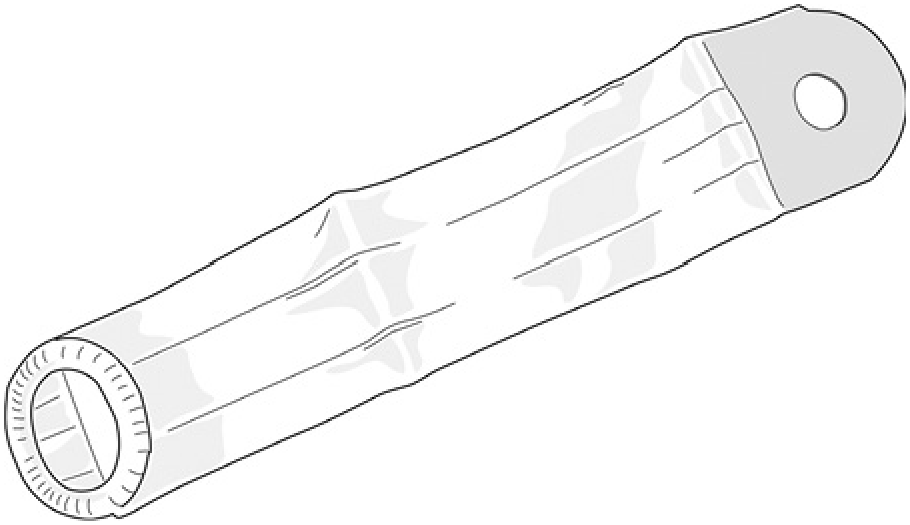


26.11

The Silastic silo is covered with povidone-iodine ointment. A dry 10 cm gauze roll is wrapped around the silo and then circumferentially around the infant's abdomen to act as a protective dressing and provide support to the Silastic silo at the fascial level.

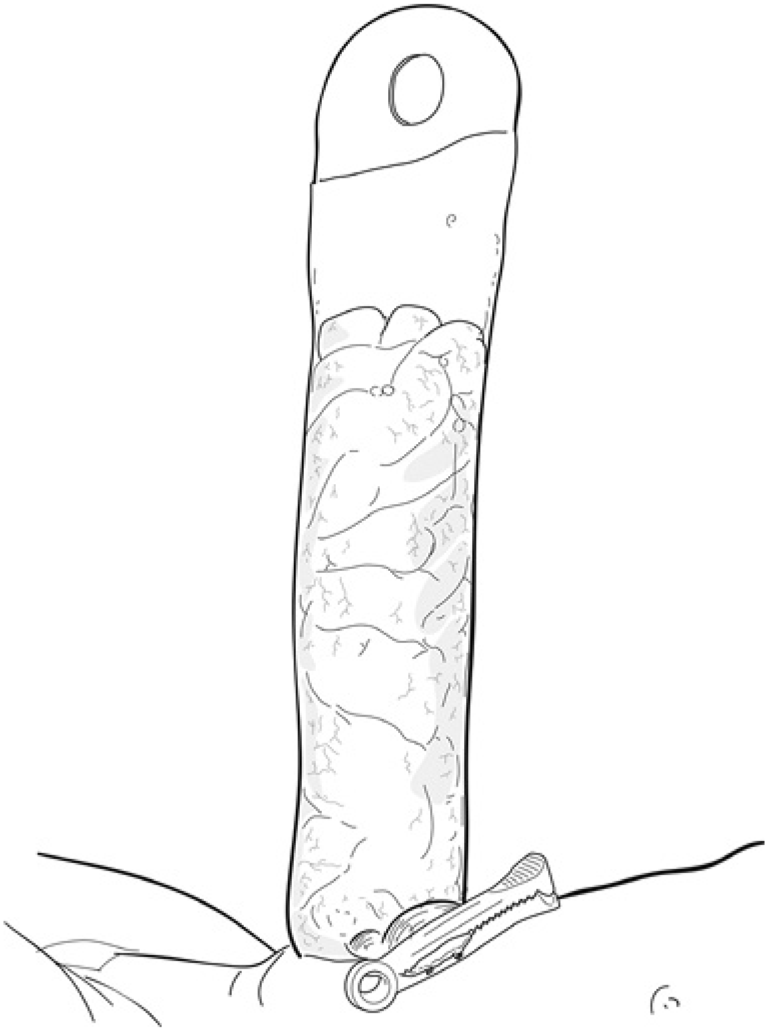
The staged reduction technique requires daily reduction of the herniated intestine within the silo. The ideal target for completely reducing the bowel, removing the Silastic silo, and closing the abdominal wall is within 1 week of age. Each subsequent day of delay beyond 1 week substantially increases the risk of fascial infection, tearing away of the Silastic silo from the anterior abdominal wall fascia, and failure of the technique. Daily reduction of the intestinal contents within the silo can be accomplished in the neonatal intensive care unit using sedation and sterile technique. Each time the procedure is performed, the silo and anterior abdominal wall are prepared with warm povidone-iodine solution before the reduction, and povidone-iodine ointment is applied followed by replacement of the gauze roll after the procedure. General anesthesia is not necessary. When the herniated bowel has been successfully reduced into the peritoneal cavity and the fascial edges brought to within 1 cm of each other, the infant is ready for removal of the silo and primary abdominal wall closure in the operating room under general anesthesia.

An alternative method is the placement of a preformed, spring-loaded silo at the bedside (**Figure 26.12**). Specific to gastroschisis, this approach has become the procedure of choice if complete bowel reduction and primary abdominal wall closure are not possible. Placement of the spring-loaded silo can be accomplished without general anesthesia. This device comes in different diameters and should be selected appropriately to accommodate the size of the defect and bulkiness of the herniated contents.



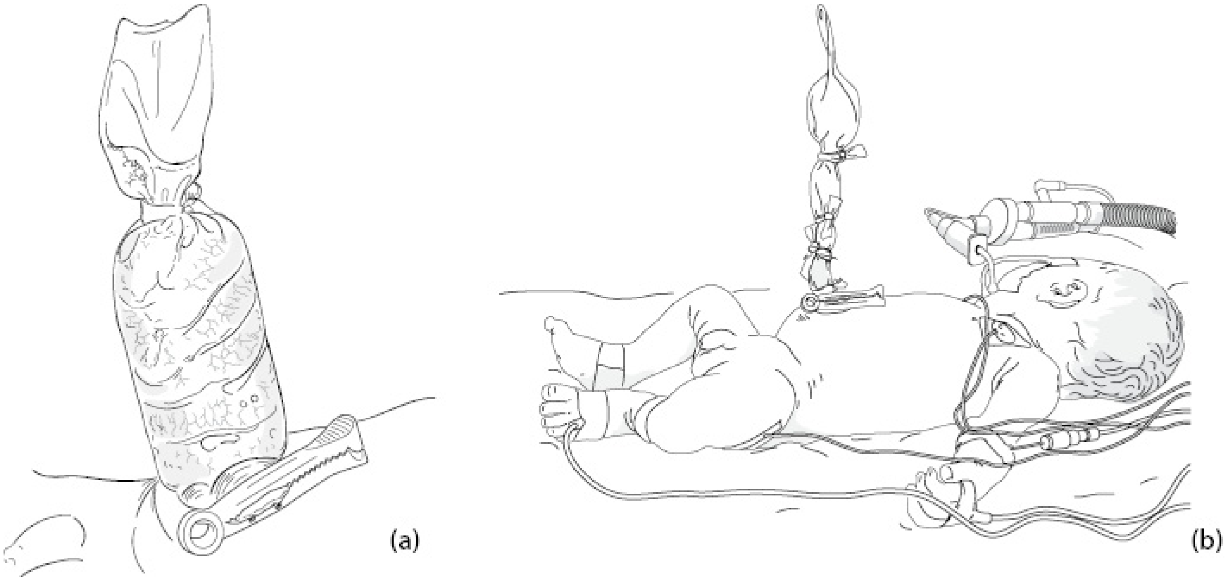
26.12

The spring-loaded silo is placed underneath the fascial defect after the herniated bowel is placed within it (**Figure 26.13**). A very small fascial defect can be constrictive and may lead to intestinal ischemia and perforation. If this is the case, the fascial opening should be enlarged, and a larger preformed silo selected prior to silo placement to prevent this potential complication.



26.13

Reduction of the bowel is accomplished in a similar fashion to that used for the sutured silo, except that, after reducing more bowel into the peritoneal cavity, an umbilical tape is tied around the silo just above the level of the intestine (**Figure 26.14a and b**).



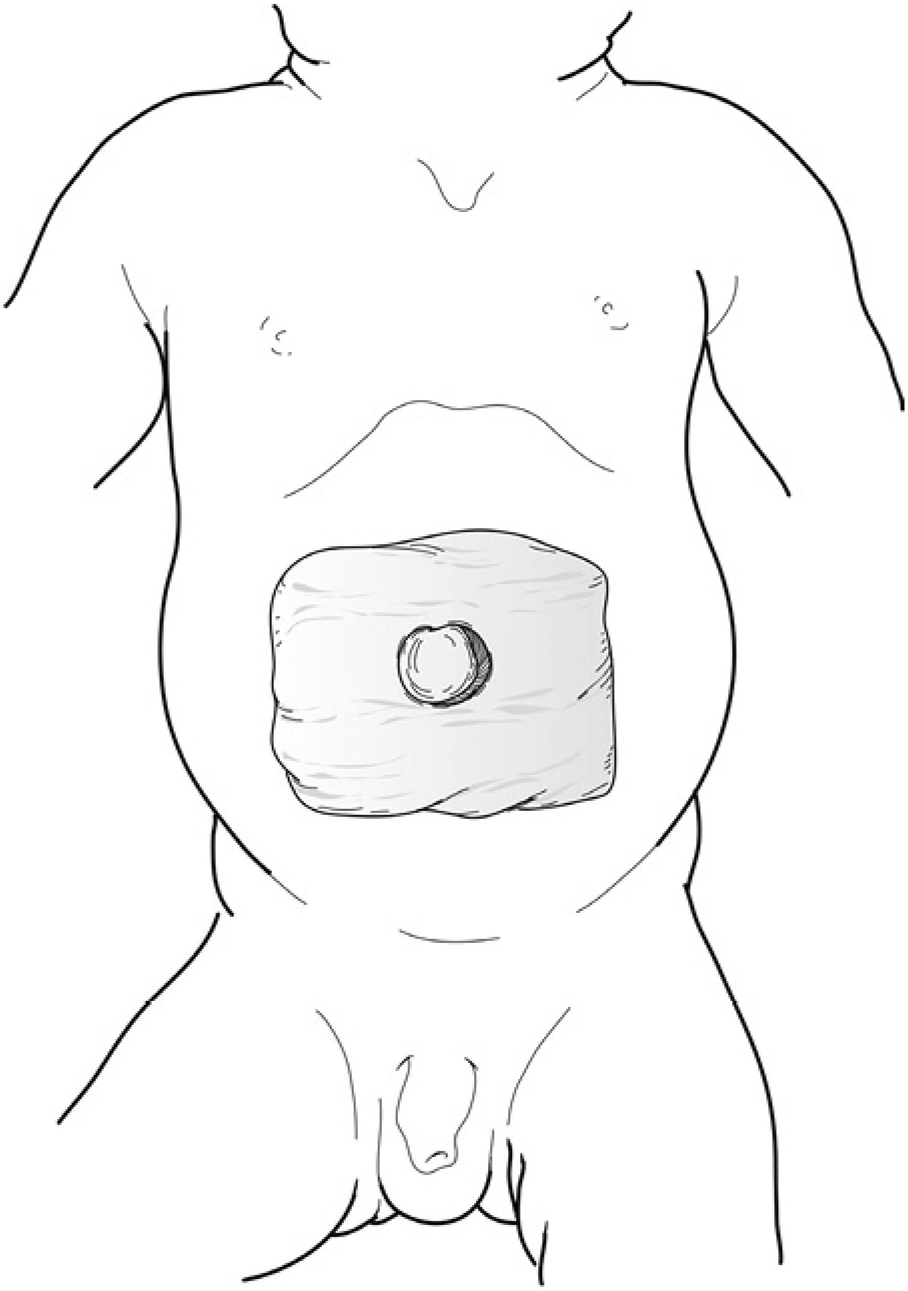
26.14a,b

As previously noted, the umbilicus is usually left in place because it results in a more normal-looking anterior abdomen. If for some reason the umbilicus has been removed, a purse-string suture can be placed in the circular skin defect subcuticularly and tied. **Figure 26.15** shows the appearance after fascial closure and purse-string technique of skin closure where the umbilicus was removed.

26.15

An alternative "gentle touch" technique has also been described. This method involves bedside placement of a spring-loaded silo, followed by passive reduction of the herniated contents into the abdomen via gravity. The infant is kept paralyzed or sedated and intubated with assisted ventilation during the passive reduction. The author states that this process typically takes 4–5 days. Subsequently, closure of the abdominal wall defect is performed.

Recently, a new method for "sutureless" gastroschisis closure has been described. In this technique, the bowel is reduced in the usual fashion either primarily or after placement in a silo. However, instead of placing sutures to approximate the fascia, the defect is covered with the umbilical stump or a non-adherent dressing (**Figure 26.16**). Occlusive dressings are then placed over the site and the wound is allowed to granulate. Once granulation tissue covers the wound bed, the area is covered with dry dressings. Proposed advantages of this technique include insignificant changes in intra-abdominal pressure and decreased narcotic and sedation requirements. Nearly all infants have an umbilical hernia following this method of repair, but many of these resolve spontaneously, similar to isolated umbilical hernias.



26.16

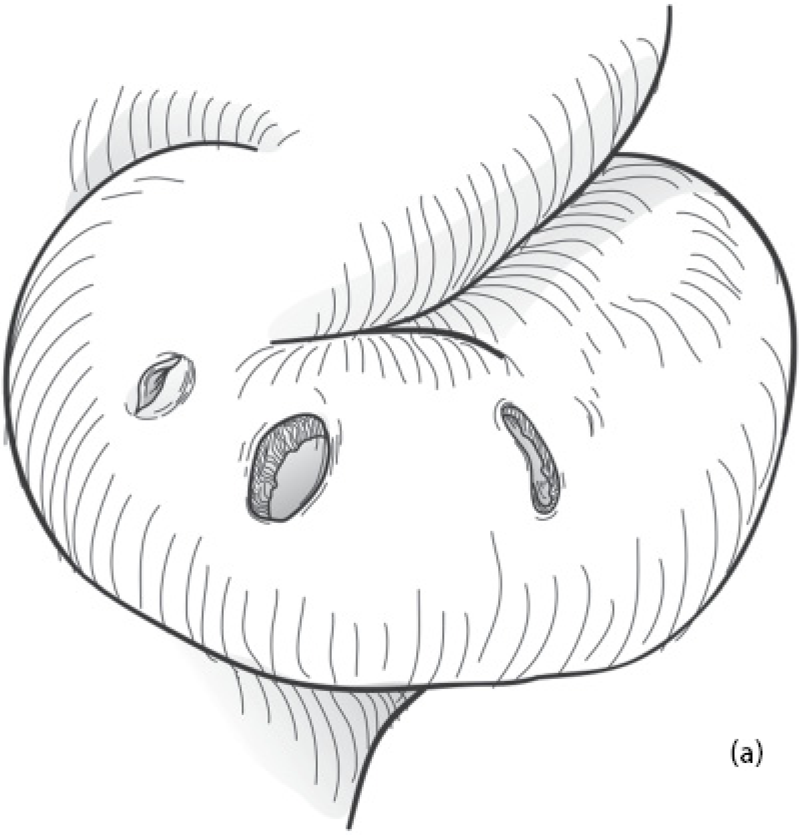
POSTOPERATIVE CARE

Most patients with gastroschisis require parenteral nutrition to provide the necessary calories intravenously while awaiting bowel reduction and recovery of bowel function. Until recently, this was accomplished via a cuffed Silastic central venous catheter. An alternative is the placement of a small Silastic catheter inserted via a peripheral vein and threaded centrally (commonly referred to as a PICC line). Parenteral nutrition is typically required for 2–4 weeks after the operation while awaiting the return of intestinal function. Nasogastric decompression is necessary until there is evidence of bowel function. Broad-spectrum antibiotics are generally continued for a minimum of 5 days. Those infants who undergo the staged approach require a longer period of antibiotic treatment (usually until 1–2 days after the silo has been removed).

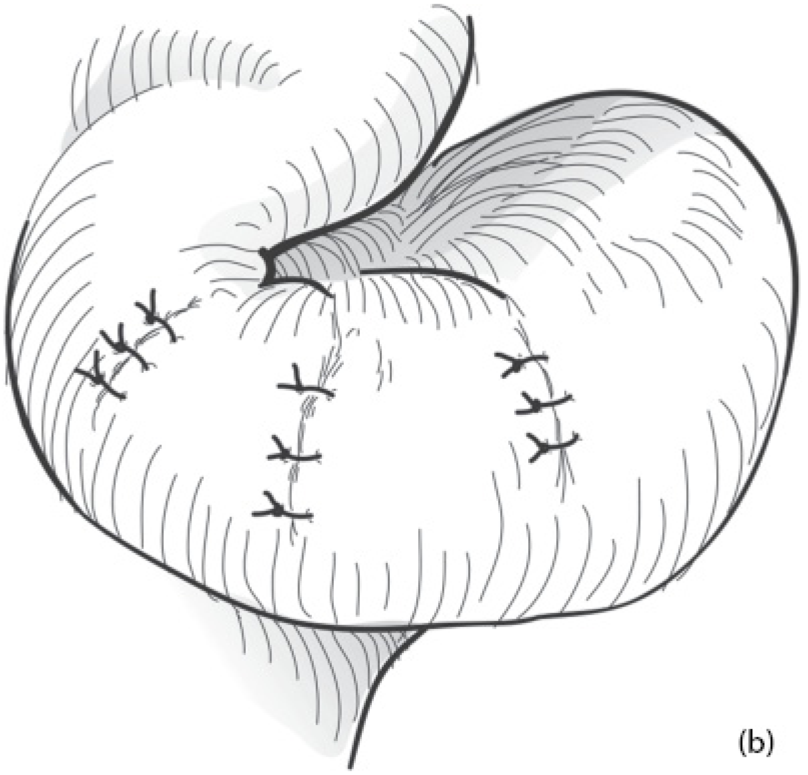
Once there is evidence of gastrointestinal function, enteral feeding can be introduced and gradually progressed using breast milk or a low-residue elemental-type formula with appropriate caloric intake by body weight. In the past, enteral feeding in these patients was generally delayed for at least 4–6 weeks after surgery, as it was thought that early feeding could lead to an increased risk of developing complications. However, this approach has not been supported with clinical evidence and, more recently, enteral feedings have been started as early as 7–14 days postoperatively, with no increase in adverse outcomes.

COMPLICATIONS

Complications in infants with gastroschisis are generally related to the gastrointestinal tract or the abdominal wall closure. As noted earlier, *in utero* complications from intestinal atresia or perforation can occur. Intestinal perforation can be managed in one of several ways, depending on the specific circumstances. The options at the time of birth include suture closure (**Figure 26.17a and b**), resection of the site of perforation with oversewing of the two ends of the bowel (i.e. creating “intestinal atresia”), or creation of a stoma if primary abdominal wall closure can be accomplished.



(a)



(b)

26.17a,b

It is generally not recommended to attempt a bowel resection with anastomosis because of the marked thickening and inflammation of the intestinal wall. Intestinal atresia can be delayed by the creation of a stoma if primary abdominal wall closure is possible or by delaying the atresia repair if staged reduction is undertaken. A stoma can be created at the time of removal of the Silastic silo and primary abdominal wall closure. Another and devastating *in utero* complication can be loss of midgut blood supply by volvulus or kinking at the level of the abdominal wall defect, resulting in midgut infarction and being born with a short segment of jejunum and residual transverse and left colon. These unfortunate infants will have extreme short bowel syndrome.

Another source of intestinal infarction can occur postnatally because of excessive intra-abdominal pressure or kinking of the blood supply to the bowel at the time of reduction of the herniated bowel. This complication may lead to the death of the patient or to short bowel syndrome.

Additional complications associated with the abdominal wall closure are wound dehiscence and intestinal–cutaneous fistula formation. These complications are also often associated with excessive intra-abdominal pressure and excessive tension on the abdominal wall closure. It is preferable to use the staged reduction approach when primary abdominal wall closure might result in excessive intra-abdominal pressure. Wound infection is relatively common, especially following delayed abdominal wall closure due to colonization of the wound from the presence of the foreign body (silo).

A delayed complication is the development of necrotizing enterocolitis. The incidence of necrotizing enterocolitis in patients with gastroschisis has been reported to be as high as 20%. It generally has a delayed onset, usually 3–6 weeks after birth. The cause remains unknown, but associations have been made with total parenteral nutrition (TPN)-induced cholestasis, excessive or accelerated postoperative feeding. Necrotizing enterocolitis associated with gastroschisis can be mild or severe and can involve a significant portion of the bowel, resulting in a high mortality.

Finally, sepsis, resulting from intra-abdominal or wound infections, and central line infections are additional causes of morbidity in patients with gastroschisis.

OUTCOME

The availability of neonatal intensive care units, parenteral nutrition, and the technique of staged reduction have resulted in significant improvement in the outcome for infants with gastroschisis over the past four decades. The survival of infants with gastroschisis has exceeded 90%. Morbidity should be relatively low if attention is paid to the details of the surgical correction. In the authors' experience, infants successfully treated for gastroschisis do not have significant complications during later infancy and childhood.

In addition to the marked improvement in survival, the length of time to initiation of feeding and hospital discharge has been significantly shortened. The average length of hospitalization is now around 4 weeks.

FURTHER READING

Badillo AT, Hedrick HL, Wilson RD, et al. Prenatal ultrasonographic gastrointestinal abnormalities in fetuses with gastroschisis do not correlate with postnatal outcomes. *J Pediatr Surg* 2007; 43: 647–53.

Jona JZ. The "gentle touch" technique in the treatment of gastroschisis. *J Pediatr Surg* 2003; 38: 1036–8.

Lacey SR, Carris LA, Beyer AJ 3rd, Azizkhan RG. Bladder pressure monitoring significantly enhances care of infants with abdominal wall defects: A prospective clinical study. *J Pediatr Surg* 1993; 28: 1370–5.

Oldham KT, Coran AG, Drongowski RA, et al. The development of necrotizing enterocolitis following repair of gastroschisis: A surprisingly high incidence. *J Pediatr Surg* 1988; 23: 945–9.

Riboh J, Abrajano CT, Garber K, et al. Outcomes of sutureless gastroschisis closure. *J Pediatr Surg* 2009; 44: 1947–51.

Schlatter M, Norris K, Uitvlugt N, et al. Improved outcomes in the treatment of gastroschisis using a preformed silo and delayed repair approach. *J Pediatr Surg* 2003; 38: 459–64.

Schuster SR. A new method for the staged repair of large omphaloceles. *Surg Gynecol Obstet* 1967; 125: 837–50.

Schwartz MZ, Tyson KR, Milliorn K, Lobe TE. Staged reduction using a Silastic sac is the treatment of choice for large congenital abdominal wall defects. *J Pediatr Surg* 1983; 18: 713–19.

27

Abdominal surgery: General principles of access

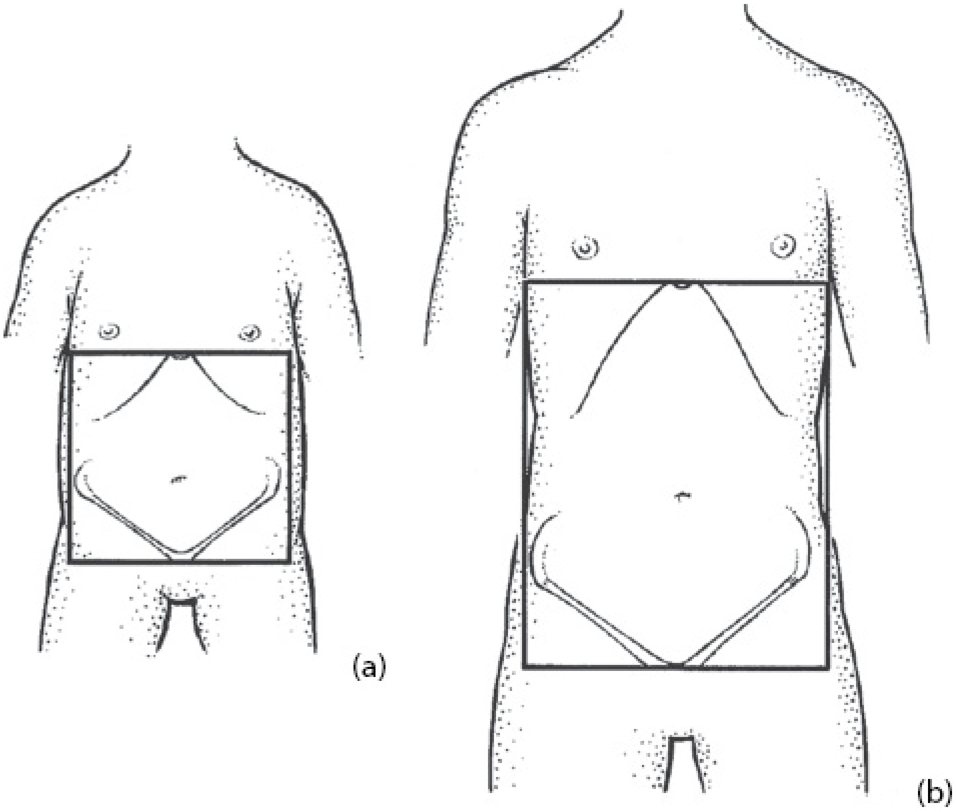
NIGEL J. HALL AND KATHERINE A. BARSNESS

OPEN SURGERY

PRINCIPLES AND JUSTIFICATION

There is no “one-size-fits-all” approach for access and we encourage selection of an incision best suited for the procedure being performed, considering the size and shape of the individual child and given the technical ability of the surgeon. In this chapter we consider the general principles of abdominal access rather than providing a recommended incision for each individual procedure since these are covered in the relevant chapter.

There are anatomic differences between the abdomen of the newborn infant and that of the older child (**Figure 27.1a and b**). When compared to an older child, an infant has a shorter, more square-shaped abdomen, with a relatively compliant ribcage and wide subcostal angle. An infant’s liver typically extends further below the costal margin, the rectus muscle is wide, extending laterally from the midline, and the umbilicus is relatively low. As a result, a supraumbilical transverse incision can provide excellent access to most of the abdomen in an infant, including the diaphragm, perhaps with exception of the pelvis. Many surgeons consider this the default incision for laparotomy in an infant or younger child. An additional benefit is that a transverse incision crosses fewer dermatomes than a midline or other longitudinal incision, resulting in reduced analgesia requirement. Particular care should be taken, however, in the preterm infant since the liver is large compared to the size of the abdomen and can extend to the level of the umbilicus and occasionally lower, and also well across the midline to the right. Some surgeons recommend a lower, infraumbilical incision in such babies.



27.1a,b

In general terms, the older the child (and therefore the larger the abdomen), the more specific the site of the incision can be, tailored for the individual procedure being performed. Clearly, the operative findings cannot be anticipated with precision in all cases so for a "utility incision" many surgeons will continue to use a supraumbilical transverse incision for a pre-pubertal child but a midline laparotomy for a post-pubertal child, with a longer more rectangular abdominal cavity.

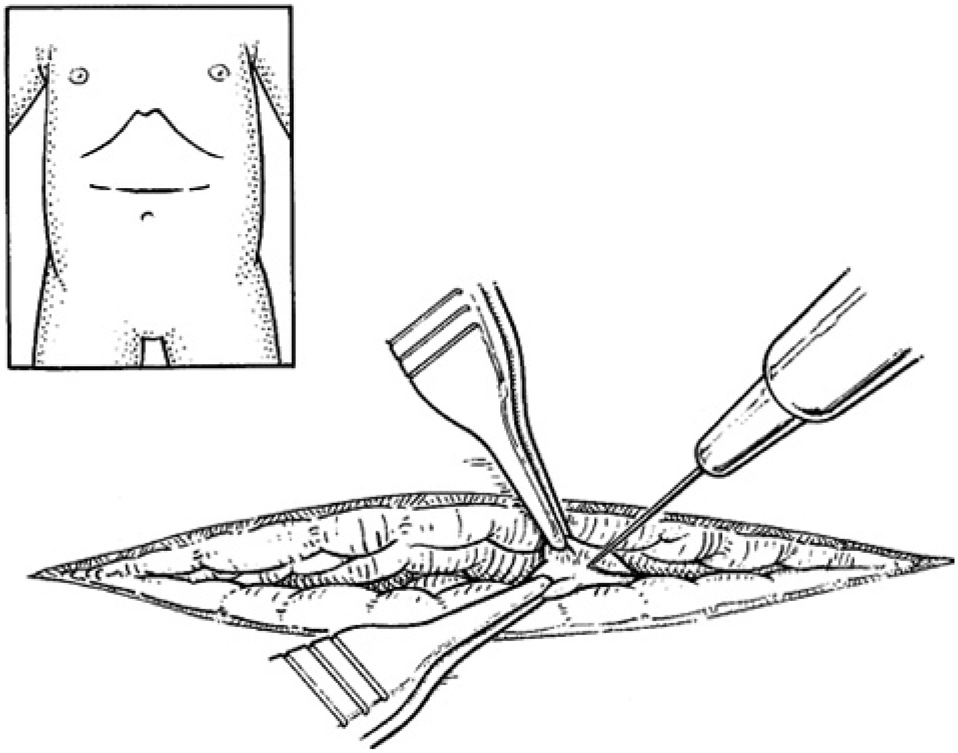
The length of the incision should be considered from the outset and kept to the minimum required to perform the procedure safely and effectively. A short incision can always be lengthened whereas a long incision cannot be shortened. Shorter incisions typically involve less cutting of muscle and therefore reduced surgical trauma. If an incision is correctly sited, it does not need to be unduly long. There is no doubt that the adoption of minimally invasive surgery in pediatric surgery has encouraged surgeons to consider how the benefits of a minimally invasive approach can be incorporated into open surgery. As a result, there is rightly a greater emphasis on a shorter incision and reduced surgical trauma and greater importance placed on cosmesis after open surgery than was previously the case. Indeed, a specific incision has been created precisely for this purpose – the periumbilical incision – through which a full laparotomy may be performed in some circumstances.

INCISIONS

Transverse abdominal incision

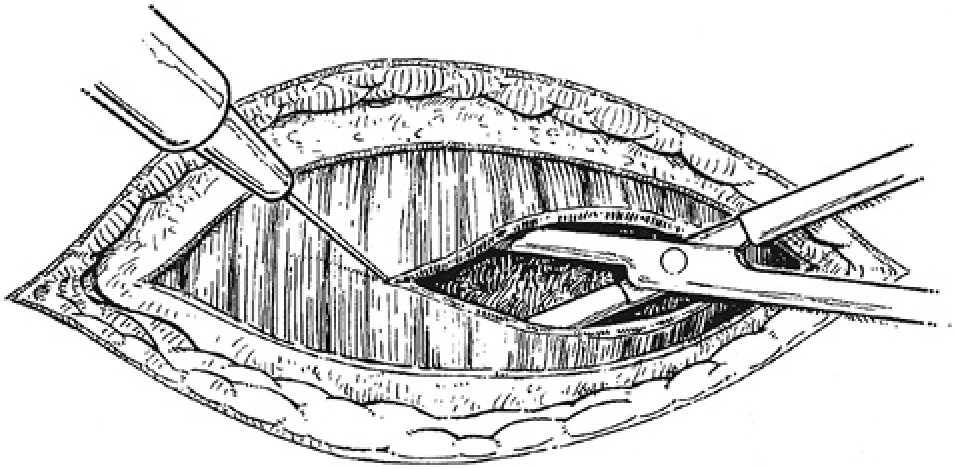
The muscle-cutting transverse upper abdominal incision is suitable for most operations in infants, except when access is required to the distal colon and rectum or other pelvic organs. The incision may be limited to one side of the abdomen or can be extended across the midline, dividing both rectus muscles. For some procedures, such as reduction of an intussusception, a transverse incision lateral to the right rectus muscle will suffice. For the emergency laparotomy when operative findings are unknown, we encourage surgeons to start with a small central incision, considering the need for lateral extension as the procedure progresses.

With the child supine, the skin incision starts in the midline, 1–2 cm above the umbilicus, and extends laterally across the rectus muscle. The subcutaneous fat is lifted with two pairs of fine-toothed forceps (one held by the surgeon and one by the assistant) and cut with diathermy to minimize blood loss, particularly in the small infant (**Figure 27.2**). Bleeding from small vessels in the skin edge will stop spontaneously with compression; larger vessels are touch-coagulated with needlepoint diathermy or picked up accurately with fine-toothed forceps and coagulated, taking care not to damage the skin.



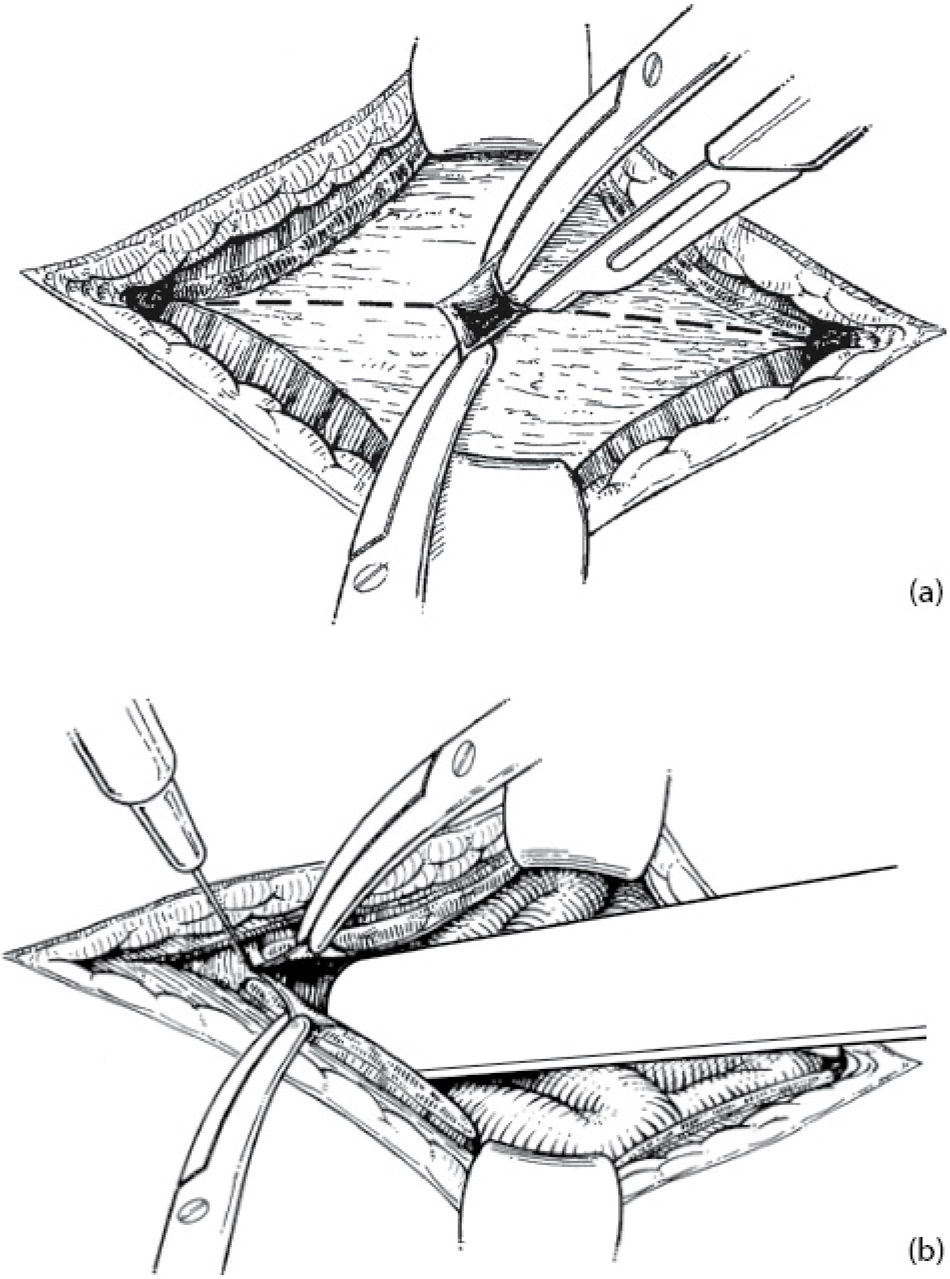
27.2

This exposes the anterior rectus sheath, which is incised transversely. A pair of artery forceps inserted deep to the rectus muscle can be used to lift the muscle off the underlying fascia while cutting it with the diathermy (**Figure 27.3**).



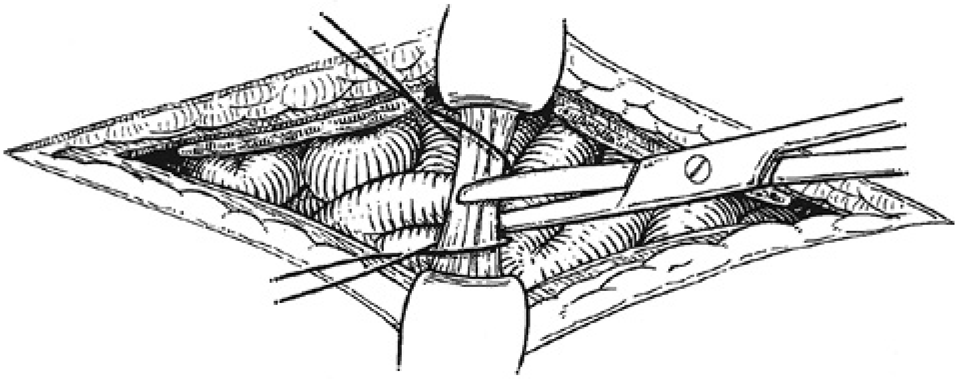
27.3

The posterior rectus sheath is picked up with two pairs of artery forceps and a small incision is made between them, taking care not to damage the underlying bowel (**Figure 27.4a**). Once air has entered the peritoneal cavity, the bowel falls away, unless it is distended or there are adhesions, and the incision can be completed safely. In the neonate, the transversalis muscle is well developed and vascular and may be divided using diathermy. The incision can be extended laterally as much as is necessary for the procedure by lifting the abdominal wall muscles with artery forceps applied to the upper and lower edges of the posterior rectus sheath, and cutting the external oblique, internal oblique, and transversalis muscle layers with the diathermy, while protecting the underlying bowel (**Figure 27.4b**). Particular care should be taken to avoid damage to distended bowel, which should be protected while completing the incision. Additional care should also be taken in cases of previous abdominal surgery or previous abdominal sepsis as there may be adhesions. These may require careful dissection from the posterior aspect of the incision when a pre-existing incision is being reopened. If possible, it is advisable to begin the incision beyond the end of the scar so that the peritoneum is opened where it is "normal" and underlying adhesions are less likely to be present.



27.4a,b

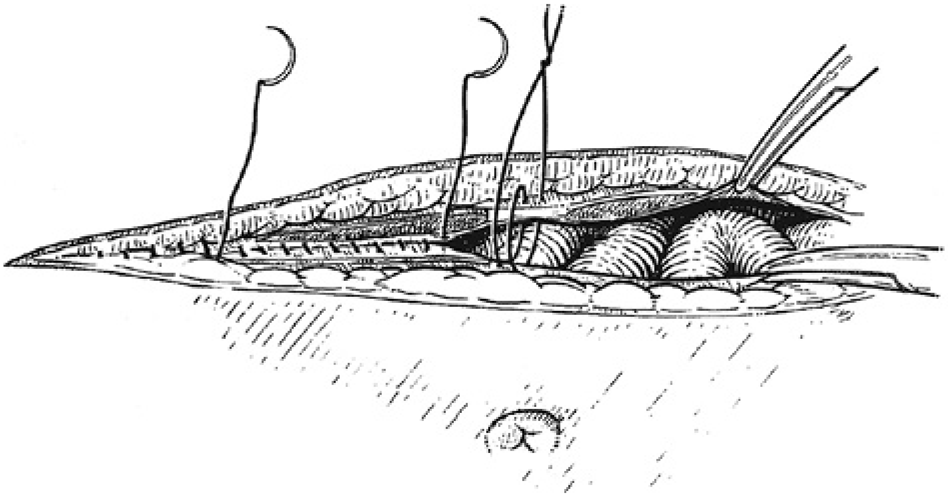
When the incision crosses the midline, the falciform ligament is encountered, containing the remnant of the umbilical vein (**Figure 27.5**). In an infant, this should be ligated and divided; in the older child, it can simply be divided.



27.5

WOUND CLOSURE

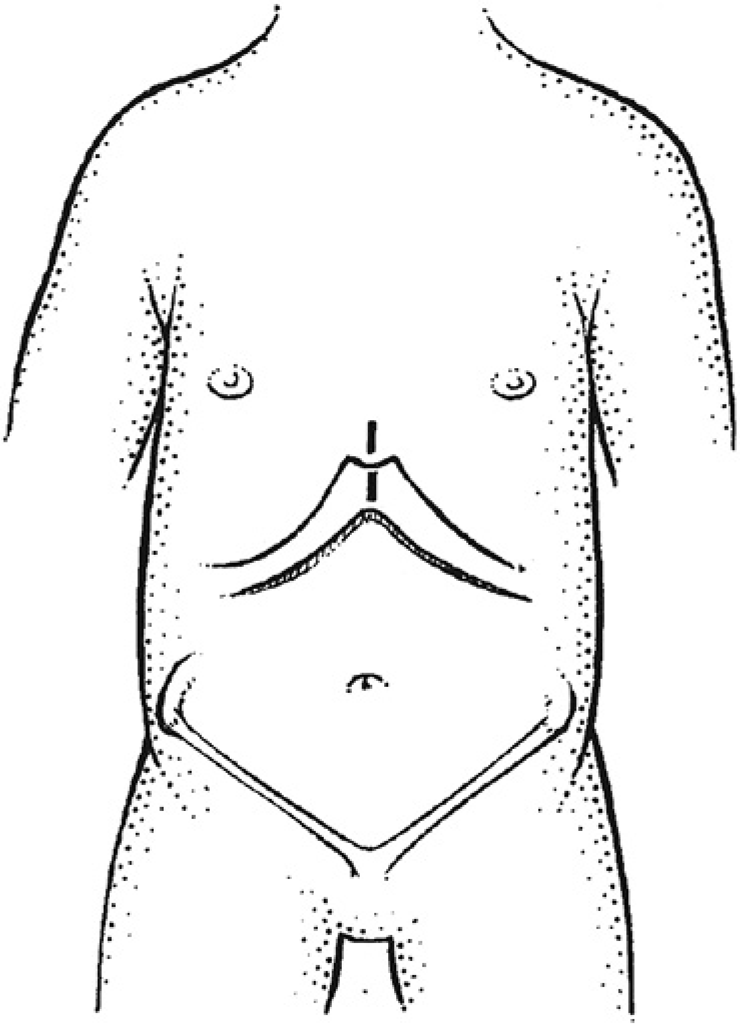
The abdominal wall is closed in layers using absorbable sutures. Interrupted or continuous sutures may be used. The principle should be to oppose each layer securely without undue tension. This may be achieved through two continuous sutures, one through the peritoneum and transversalis muscle or fascia, the second through the anterior rectus sheath (**Figure 27.6**). Alternatively, a series of interrupted figure-of-eight sutures may be placed along the entire length of the incision. If closing a pre-existing incision, it may not be possible to separate the layers of the anatomical wall, in which case a continuous "mass" closure incorporating all layers may be used. Having closed the muscle, the subcutaneous fascia is closed, followed by the skin.



27.6

Subcostal incisions

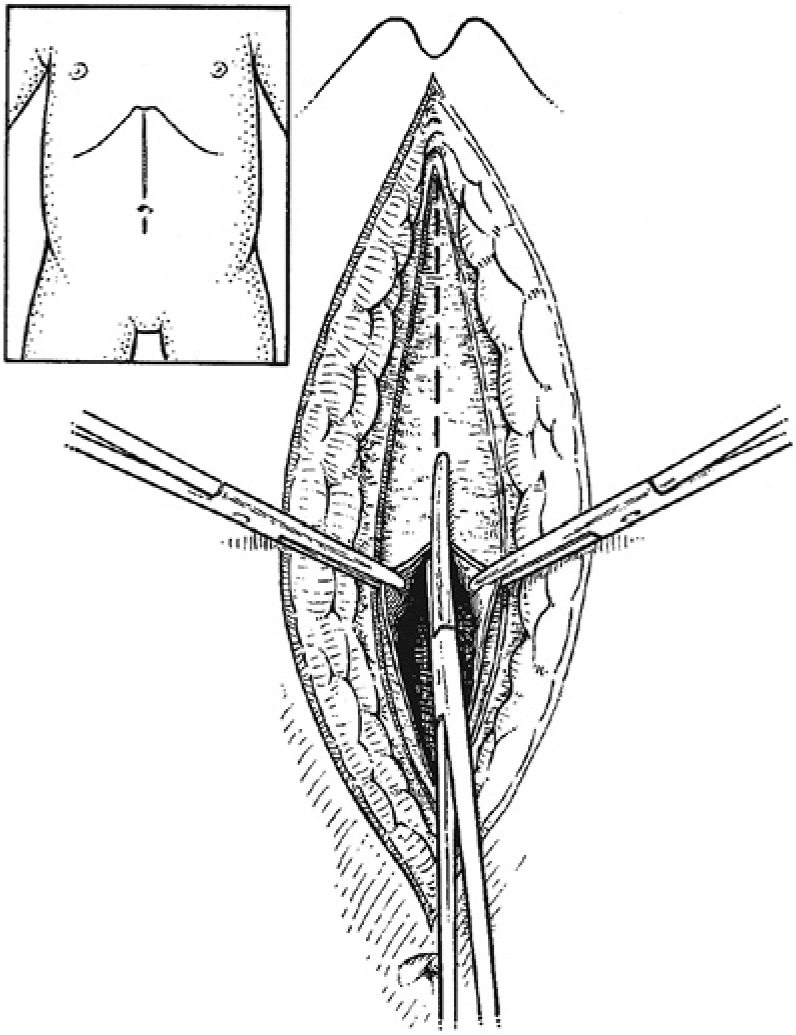
A subcostal incision and the bilateral subcostal or “rooftop” incision are both useful for access to the upper abdomen (**Figure 27.7**). The principles of incision and wound closure are the same for other abdominal muscle-cutting incisions.



27.7

Midline abdominal incision

The skin is incised in the midline over a length and location appropriate for the incision. This may be entirely supraumbilical (e.g. gastrostomy, fundoplication) entirely infraumbilical, or above and below the umbilicus, either continuing vertically through the umbilicus or curving around one side of it. Having divided subcutaneous fat and fascia, the midline linea alba is encountered and incised over a short distance with a scalpel. Above the umbilicus, the falciform ligament is entered and incised to enter the peritoneal cavity. The edges of the incision are elevated and a plane is developed deep to the linea alba, which is then incised. Below the umbilicus, the recti muscles sit behind the strong fascial layers and can be split in the midline (**Figure 27.8**). Above the pubic bone care should be taken to avoid inadvertently entering the bladder, particularly in a small child.

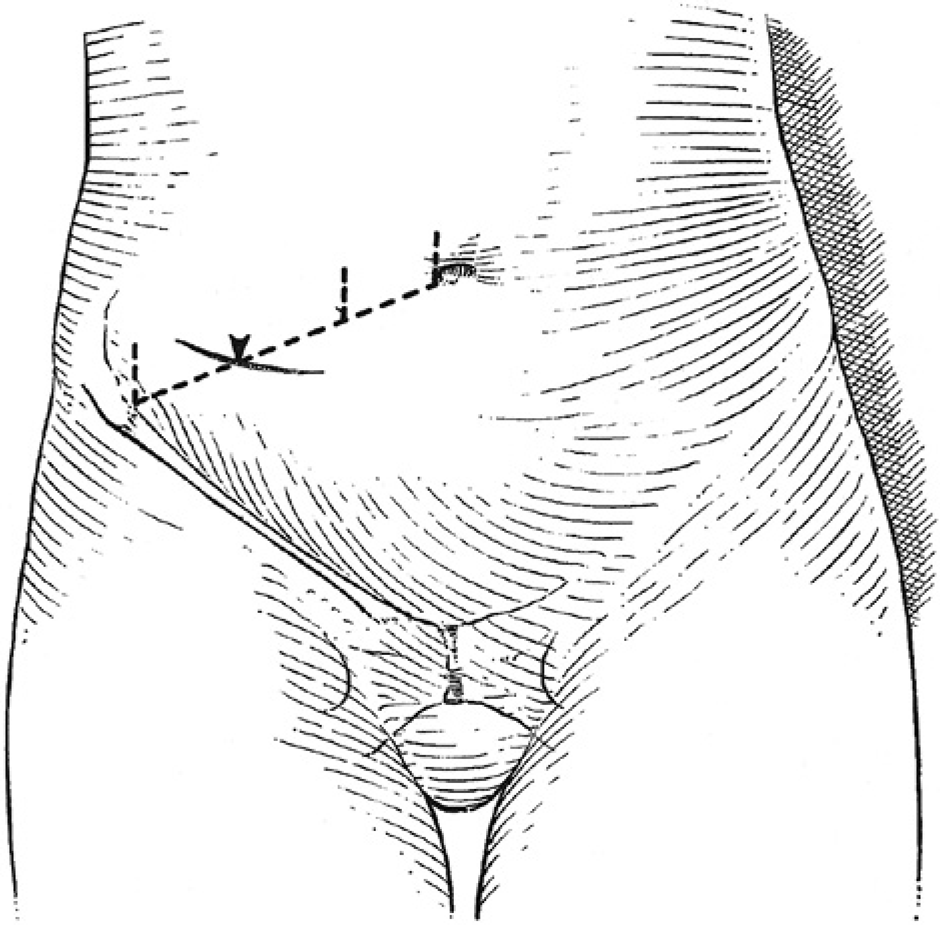


27.8

At the time of closure, the linea alba is approximated with a continuous strong slowly absorbable suture. The knot at each end can be buried to avoid an unsightly nodule prior to closing the overlying subcutaneous tissue and skin.

Grid-iron incision

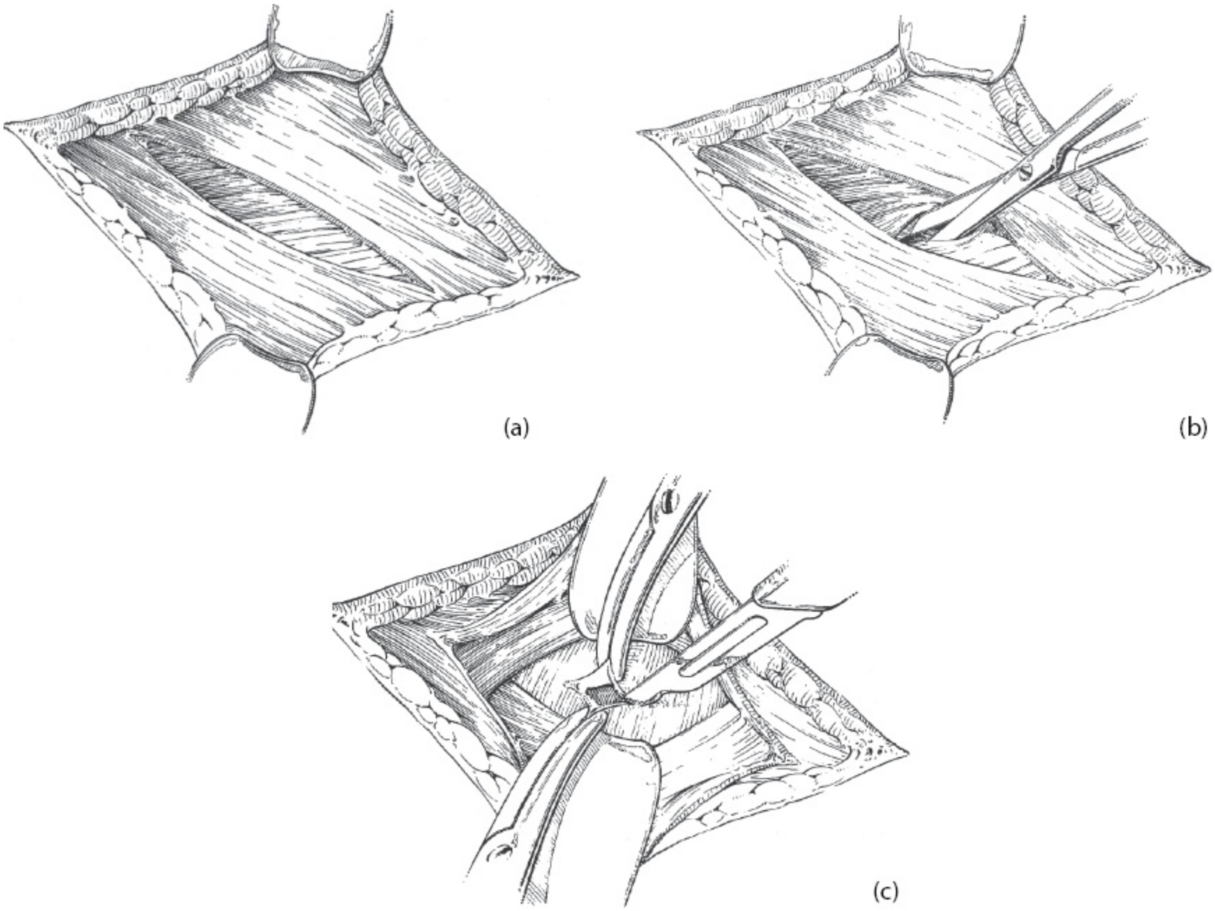
This modified McBurney* incision is the ideal incision for acute open appendectomy in childhood. The traditional incision is centered over McBurney's point, which is two-thirds of the distance from the umbilicus to the right anterior superior iliac spine, and is at right angles to this line (**Figure 27.9**). Nowadays, the skin incision is more horizontal and aligned with the skin creases and termed a Lanz incision. It should be lateral to the rectus muscle, which is relatively broad in a child. A lower incision and more transverse skin crease incision may be preferred for cosmetic reasons, but exposure of the appendix may be difficult if the incision is too low or too medial.



27.9

The subcutaneous tissues are divided with diathermy and swept aside with a swab to expose clearly the external oblique aponeurosis; this facilitates subsequent closure. The external oblique muscle is incised and then split along the line of its fibers before separating it from the underlying internal oblique muscle (**Figure 27.10a**). The internal oblique muscle is split with blunt dissection, spreading it at right angles to the direction of the fibers and completing the “split” with two Langenbeck retractors. The underlying transversus abdominis muscle and the fatty layer covering the peritoneum are opened in a similar fashion (**Figure 27.10b**).

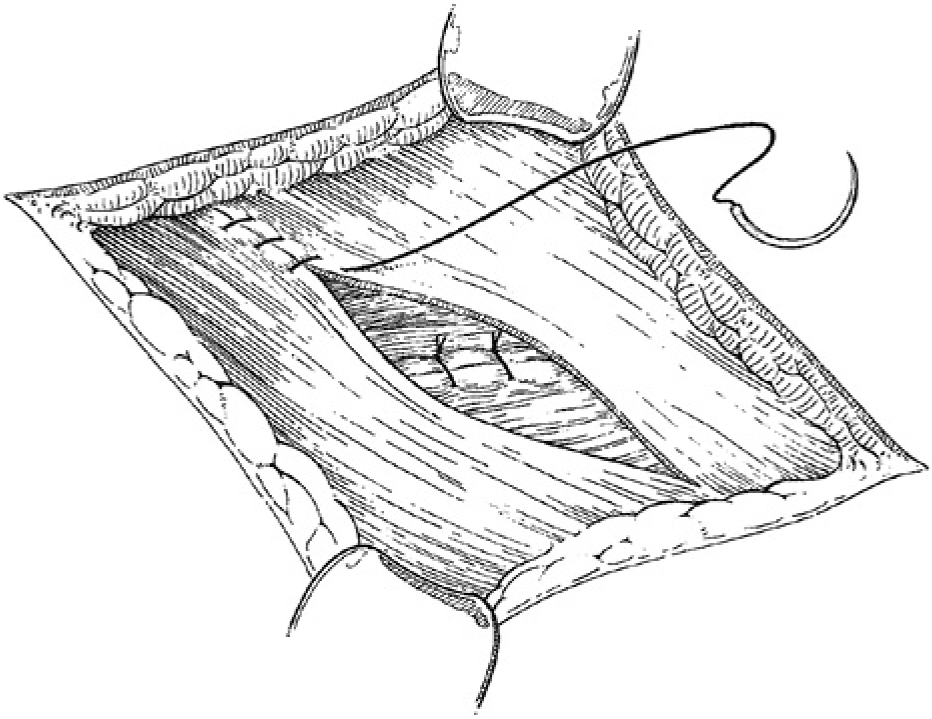
The peritoneum is grasped with two pairs of artery forceps, taking care to avoid the underlying bowel. The forceps are lifted and the peritoneum is incised transversely with a scalpel; the opening is enlarged using scissors (**Figure 27.10c**).



27.10a–c

WOUND CLOSURE

The edges of the peritoneum are grasped with artery forceps and closed with a continuous absorbable suture. The fibers of the transversus and internal oblique muscles are closed as a single layer, using two or three interrupted sutures, which are tied loosely to avoid muscle ischemia. The external oblique muscle is closed with a continuous absorbable suture (**Figure 27.11**). Scarpa's fascia followed by the skin can then be closed with an absorbable suture.



27.11

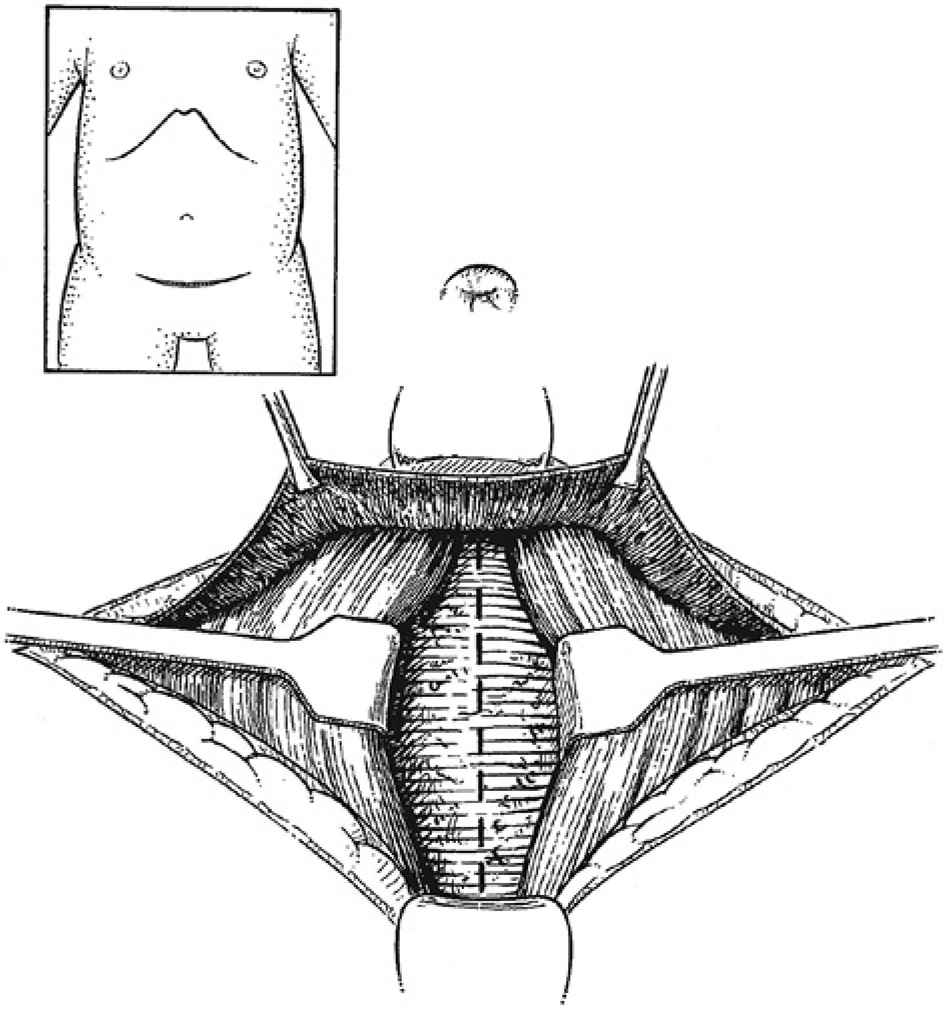
EXTENDING THE INCISION

If the incision needs to be extended laterally, the abdominal muscles can be divided in layers using the diathermy. To extend the incision medially, the incision in the external oblique muscle is continued onto the anterior rectus sheath. The rectus muscle is retracted medially. The internal oblique and transversalis muscles are divided medially and this incision is extended to open the posterior rectus sheath and peritoneum. If necessary, the rectus muscle is also divided. The incision is closed in layers, as for a transverse abdominal incision.

Pfannenstiel incision

The Pfannenstiel incision* is a lower abdominal incision that provides access to the pelvic organs, in particular the bladder, uterus, and ovaries, and results in a cosmetically acceptable scar. Depending on the degree of access required, division of the rectus muscles may be avoided completely.

The skin and subcutaneous tissues are incised transversely about 2 cm above the pubic symphysis and extended laterally as necessary, curving upward at each end. The anterior rectus sheaths are divided transversely. If the muscles are to be preserved, the anterior rectus sheath is dissected off the underlying muscles by blunt and sharp dissection, extending cranially well up to the umbilicus and caudally to the pubic symphysis. The rectus muscles are separated vertically in the midline and retracted laterally (**Figure 27.12**). The transversalis fascia and peritoneum are then opened vertically, taking care to avoid the bladder. If a greater degree of pelvic access is required, the lower rectus muscles are divided transversely rather than separated following the principles described above.

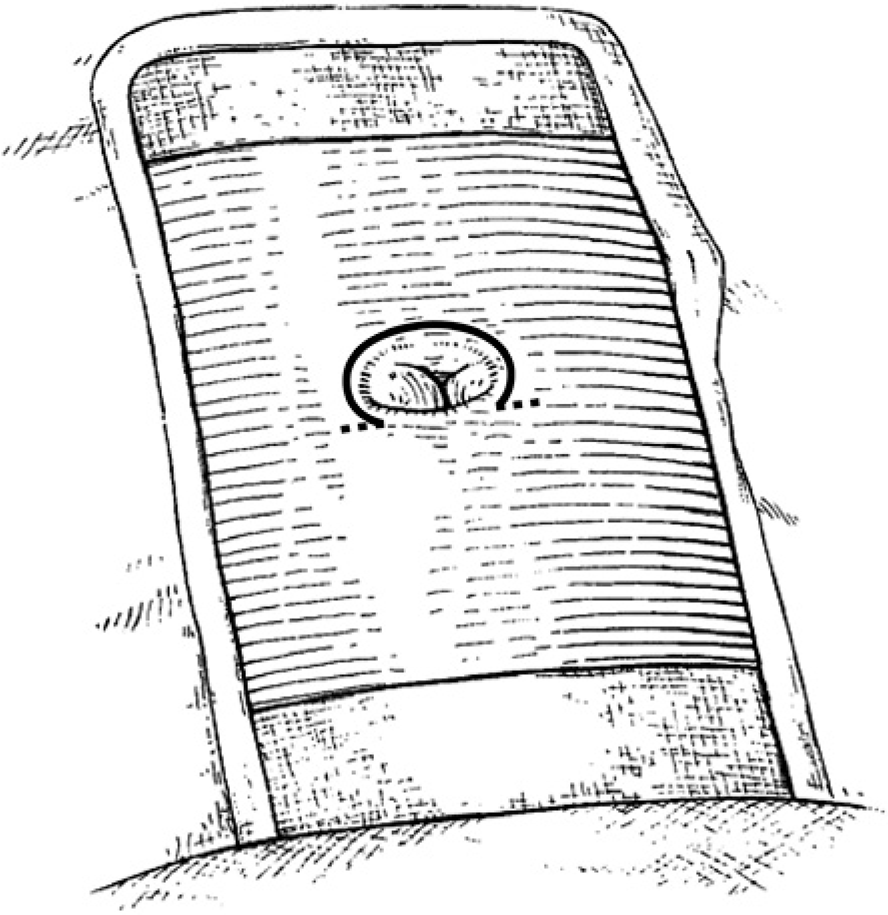


27.12

Closing the vertically separated muscles simply involves approximation in the midline with interrupted absorbable sutures.

Periumbilical incisions

Over the past 20 years it has been increasingly realized that a periumbilical approach can be utilized for a number of abdominal surgical procedures, particularly in infants, to provide access to the abdominal cavity, yet leaving very little in the way of a surgical scar. Indeed the "natural scar" of the umbilicus is in some ways ideal for this purpose. The supraumbilical approach to pyloromyotomy as proposed initially by Adrian Bianchi has become the preferred open approach to this operation in most centers as it provides vastly superior cosmesis to the midline or transverse right upper quadrant incisions. By modifying this, a relatively long incision, providing adequate access for many intestinal procedures in infants, can be achieved. The skin around the umbilicus is incised for anything up to three-quarters of the circumference and can be extended if necessary laterally into an omega-shaped skin incision. The muscle sheath is divided transversely and the rectus muscles either retracted laterally or similarly divided transversely (**Figure 27.13**). The umbilical vein is divided or retracted and the intestine delivered. Repair of duodenal and intestinal atresia, correction of malrotation, excision of Meckel's diverticulum, as well as other procedures have all been reported using this approach.



27.13

On closure, the muscle layer is repaired and the skin incision closed. If an omega incision has been created, the lateral triangles can be excised to leave a curved scar which blends inconspicuously with the umbilicus itself.

LAPAROSCOPIC SURGERY

PRINCIPLES AND JUSTIFICATION

Laparoscopy is best understood as an alternate technique by which a surgeon can perform the same operation as is performed through an open incision. The principles and goals of an abdominal operation remain unchanged by the means by which the pathology or anomaly is accessed and corrected.

Laparoscopy is unique in its approach to the abdominal cavity. Whereas open incisions are placed as close to the target pathology as possible, laparoscopic port sites are better sited distant from the target, creating a wide view of the surrounding tissues and structures. The result is that laparoscopic access can provide a much larger field of view than an open incision, particularly in the absence of intra-abdominal adhesions and scarring or large space-occupying tumors. One caveat to the use of laparoscopy is the requirement that intra-abdominal domain is not compromised by the pathologic state for which the operation is being conducted. A distended abdomen that is taut with bowel dilatation or a large space-occupying lesion may prevent further distention of the abdominal wall with carbon dioxide insufflation, abrogating the ability to create a working space for a laparoscopic procedure.

Potential advantages of laparoscopy, compared to open abdominal access, include less scarring, less pain and a reduced need for opioid pain medications, reduced length of stay, and faster return of bowel function and return to normal activities. Many of these advantages are achieved by stressing small muscle-splitting incisions, in addition to the lack of exposure of the bowel to air. Additional advantages of laparoscopy include magnification of the target anatomy and the ability for all members of the surgical team to visualize the procedure, thereby encouraging greater engagement of operating room personnel for the duration of the procedure. The improved visualization is particularly relevant in neonatal disease processes when open incisions are limited by the small size of the infant, thereby limiting simultaneous visualization of the target anatomy by both the primary and assistant surgeons.

INITIAL ACCESS

Initial access into the peritoneal cavity is performed via a dissection (i.e. Hasson technique) or a needle insufflation (i.e. Veress needle) technique. Alternatively, a hybrid version of these two methods can be performed. While there are theoretical advantages to all of these approaches to obtain initial access, numerous literature reviews have failed to find an advantage of using one technique over another.

Hasson technique

The Hasson technique[*] for initial access to the peritoneal cavity is a controlled access under direct visualization of the linea alba in the midline of the abdomen. After incision in the skin, the soft tissues are dissected down to the fascia. The fascia is grasped between two instruments, and a vertical incision is made under direct visualization. A hemostat or other dissecting instrument is advanced through the incision and gentle spreading is used to create a fascial defect that approximates the size of the trocar. A blunt obturator is advanced into the trocar, and then both the obturator and trocar are advanced into the peritoneal cavity under direct visualization. Any trocar placed using a Hasson technique usually requires some type of suture stabilization of the trocar, to prevent it from falling out of the peritoneal cavity. Alternatively, some trocars have a deployable stabilizer beneath the fascia or a radially expanding sheath to stabilize the trocar, eliminating the need for a stabilizing suture.

Veress needle technique

The concept of the Veress needle[*] is that insufflation of the peritoneal cavity to a preset maximum pressure creates a safe space into which a trocar can then be inserted. While conceptually appealing, visceral injuries are equally as likely with Veress needle techniques as they are with other methods for initial and subsequent trocar placement. It is therefore advised that the method of access be done with the utmost care required, with the acknowledgment of the ever-present risk of injury to abdominal viscera from any means of laparoscopic access.

The Veress needle is a hollow, beveled needle with a spring-loaded inner cannula. The inner cannula retracts as the needle passes through muscle and fascia, but then the round-tipped inner cannula springs out beyond the sharp tip once entry into the peritoneal cavity is obtained. The inner cannula deployment is both heard and felt by the surgeon placing the Veress needle, once it passes through the fascia and there is no longer soft tissue and fascial resistance. Once intra-abdominal position is thought to have occurred, there are several methods to assess for correct positioning of the Veress needle in the peritoneal cavity. A saline test involves several steps to confirm intraperitoneal Veress needle placement. First, a 5 mL syringe filled with saline is used to aspirate to evaluate for intestinal contents, blood, or urine. If there is a negative aspiration, 5 mL of saline is gently instilled. The saline should pass easily through the Veress needle with no resistance. The syringe is then removed, leaving a drop of water on the end of the needle. The fascia is then lifted using a penetrating clip or Kocher clamp. With elevation of the fascia, the saline drop should promptly pass through the Veress needle into the peritoneal cavity.

An alternate method to evaluate for intra-abdominal position of the Veress needle is to do a blind sweep of the needle 360 degrees around the access site. In patients with previous abdominal operations or space-occupying lesions, this method is not advised, due to the risk of injury to other structures. Initiation of insufflation is the final assessment for intraperitoneal placement. With insufflation, the insufflator-measured intra-abdominal pressures should start low, with a continuous flow of carbon dioxide until the level of preset maximum abdominal insufflation pressure has been achieved.

While one or more of these methods may be useful to confirm intraperitoneal Veress needle position, none is 100% sensitive and specific. It is only with subsequent trocar placement and advancement of the telescope into the peritoneal cavity that peritoneal access can be confirmed.

Trocar placement after Veress needle insufflation of the peritoneal cavity is variable, dependent upon the type of trocar being used for the procedure. Trocars can have beveled cutting tips and inner obturators or rounded non-cutting tips and obturators. A beveled cutting tip or obturator usually does not require a fascial incision to pass into the peritoneal cavity. If the cutting trocar is the first trocar being placed, then the abdominal wall is stabilized by a penetrating clip or Kocher in the non-dominant hand, while the dominant hand is used to slowly advance the trocar through the fascia. A twisting motion of the hand is often used to assist in the passage of a cutting trocar. If a non-cutting trocar is being used, it usually has a rounded obturator for the inner cannula. The fascia is stabilized with a penetrating clamp or Kocher clamp and a small retractor is used to retract the soft tissues to allow direct visualization of the fascia. An incision is then made in the fascia, followed by advancement and spreading of the tissues with a hemostat. Once air is heard escaping from the incision with the hemostat, the trocar with its obturator in place is passed into the peritoneal space, as in the Hasson technique.

Radially expanding sheathed Veress needle technique

Radially expanding sheathed Veress needle trocars allow for a hybrid technique to be used for the placement of the initial trocar. In this trocar system there is a radially expanding sheath into which the Veress needle is initially inserted. The sheathed Veress needle is placed into the abdomen either through a Hasson method of direct visualization and incision of the fascia, or via a standard stabilized puncture of the fascia with the sheathed Veress needle. In infants, the Veress needle is removed after the sheath has passed into the peritoneal cavity, usually without insufflating through the

Veress needle. The trocar is passed through the expandable sheath and then a telescope is advanced into the trocar. If direct visualization of the peritoneal cavity is confirmed, carbon dioxide insufflation is begun under direct visualization. In older children and adolescents, the sheathed Veress needle can be used in the same way as a non-sheathed Veress needle to create pneumoperitoneum up to the desired insufflation pressure. The Veress needle is removed and the trocar is then advanced through the sheath.

Optical trocar technique

Optical trocars have a clear tip at the end of the trocar through which a zero degree telescope can visualize the tissue layers as the trocar is advanced through the myofascial planes. Optical trocars are commonly used in morbidly obese patients, utilizing Palmer's point in the left upper quadrant for the initial access site (described later). The first step to optical trocar use is creation of pneumoperitoneum using a Veress needle technique. After the preset abdominal pressure has been achieved, a skin incision is made to accommodate the size of the trocar. In one optical trocar system, the distal tip of the trocar is a clear crystal with cutting blades on it. Using slow, steady pressure with a rotating motion, the port is advanced into the peritoneal space. This type of optical trocar requires significant force generation by the surgeon to get the port to pass through the myofascial layers. A second system has a trigger system that deploys a cutting knife 1 mm beyond the clear tip of the trocar. After each deployment, the knife retracts to allow the surgeon to see progress that has been made. This system requires less force for placement that other systems.

Umbilical access

The near-universal starting point for laparoscopic access to the peritoneal cavity is the umbilicus. The umbilicus represents remnant scar tissue from involution of umbilical cord structures. As such, use of the umbilicus for trocar placement allows surgeons to “hide” an incision within the folds of a natural scar. The umbilicus also is represented by a single fascial plane that, once crossed with a Veress needle or trocar, allows for efficient entry into the peritoneal space. For children with a persistent natural patency in the center of the umbilicus, incision of the fascia may not even be required.

INFANTS

Special consideration is required for umbilical access in neonates and infants less than 1 year of age. During fetal circulation, the umbilical vein drains through the ductus venosus into the left hepatic vein, near its junction with the inferior vena cava. After birth, involution of the umbilical vein forms the falciform ligament. However, the age at which the natural patency of the remnant umbilical vein fully closes is not clearly defined. A number of case reports in the medical literature, as well as case law from malpractice lawsuits, document air embolism through the umbilical vein remnant to be a highly morbid or lethal complication of laparoscopy in infants. For this reason, both supraumbilical access and direct access through the center of the umbilicus should absolutely be avoided in neonates and infants. While the vast majority of air embolisms are noted to occur in infants of less than 3 months of age, the timing of full involution of the natural patency is not known. It should also be noted that air embolisms have been documented in cases when insufflation has not yet begun, hypothesized to be secondary to pushing air into the vein during advancement of a sheathed trocar into its Veress needle-placed sheath. Air embolism has also occurred at later stages of a procedure, when a trocar is either intentionally or inadvertently pulled back in the fascia, with resultant insufflation of the umbilical vein. As there is no absolutely safe method to place a trocar near the umbilical vein, it is therefore advised that all neonate and infant trocars be placed in an infraumbilical location.

Infraumbilical port position in a neonate or infant is easily accomplished by a curvilinear incision along the lower umbilical folds, with direct visualization of the midline fascia below the umbilical ring. A vertical incision in the midline fascia will best avoid the umbilical arteries.Using a

Hasson technique or a sheathed Veress needle, trocar placement proceeds as described.

CHILDREN AND ADOLESCENTS

As previously noted, the umbilicus is an excellent position in which a scar can be easily hidden within the folds of a natural scar. The shape, size, and depth of the umbilicus all influence the choice of incision placement to accomplish ready access with minimal scarring. In a child with an umbilical hernia, the initial skin incision is usually a curvilinear incision along the folds of the umbilical skin. The peritoneum of the hernia defect is then directly incised (Hasson) or punctured (Veress needle) for trocar placement. If the umbilical defect is larger than 1 cm, consideration can be given to placing the first port below the umbilical ring, in order to stabilize the port within the midline fascia. However, this introduces a second fascial defect to the one that already exists at the umbilical ring. In children without an umbilical fascial defect, there are two options for the initial skin incision: Directly through the base of the umbilicus or via a curvilinear incision in the folds of the umbilicus. An incision through the base of the umbilicus is best utilized in children with broad, flat umbilical rings and minimal surrounding soft tissue. A curvilinear incision is better for a deep or narrow umbilicus.

Left upper quadrant (Palmer's point) access

Palmer's point is located in the left upper quadrant, 3 cm below the left costal margin in the midclavicular line. Palmer's point access is often used in patients with previous abdominal surgery, or large pelvic or lower abdominal masses. It is also useful in morbidly obese patients, as the amount of subcutaneous fat in this location is usually less than that found at the umbilicus. In adult-sized patients, this location can be used for initial access using Hasson, Veress needle, or Veress needle/optical trocar techniques. Palmer's point is not often utilized for infants or children.

The technique for access is similar to that described previously, but care should be taken to ensure full decompression of the stomach with a nasogastric or orogastric tube. Inadvertent gastric perforation is the most common complication of access at Palmer's point, followed closely by colonic perforation. As with any access location, risk of injury to adjacent peritoneal and retroperitoneal structures is always present and should be considered when choosing any location for initial access.

CARBON DIOXIDE INSUFFLATION

Abdominal muscle relaxation is required for laparoscopic procedures of any duration. Brief evaluations (e.g. laparoscopic evaluation of the contralateral processus vaginalis during open inguinal hernia repair) can be accomplished with a laryngeal mask airway. Otherwise, an endotracheal tube is required for full paralysis or deep non-paralysis anesthetics. With full relaxation of the abdominal muscles, the abdominal wall is distended with CO_2 insufflation to create the peritoneal working space for laparoscopic procedures.

Insufflation pressures are variable, depending on the age and/or size of the patient. The insufflation goal is to achieve a reasonable working space at the lowest insufflation pressures required. One complication of CO_2 insufflation is its absorption across the peritoneal surfaces into the bloodstream, with a resultant increase in end-tidal CO_2. Carbon dioxide absorption increases with higher insufflation pressures. Other complications of high insufflation pressures include decreased renal blood flow and venous return to the heart and increased peak inspiratory pressures required for ventilation. For neonates and young infants, an insufflation pressure of 6–8 mmHg is adequate to achieve sufficient abdominal domain for most procedures. An insufflation pressure of 8–10 mmHg is usually adequate for older infants and children. Most adolescents tolerate pressures of 12–14 mmHg. Patients are ideally euvolemic, to minimize complications related to renal blood flow and venous return to the heart. In children with pulmonary disease, particularly children with ventilatory concerns and baseline CO_2 retention, laparoscopic procedures need to be discussed with the anesthesiologist in detail, to ensure parameters for tolerable CO_2 levels are agreed upon before the procedure is initiated. In these children, an arterial line may be necessary for intraoperative monitoring of CO_2 levels and overall acid base status, as end-tidal CO_2 is a sampling method that is not as reliable in young children and infants. Children with congenital heart disease, in the absence of concurrent pulmonary disease, generally tolerate laparoscopic procedures well, as long as venous return is not impaired by high insufflation pressures and the intravascular volume is closely monitored. Bladder catheterization may be required in these children, as urine output is a good proxy for overall organ perfusion in these children.

Carbon dioxide flow rates are the second variable preset by the operating surgeon. Flow is the rate at which CO_2 is delivered through a Veress needle or trocar, to maintain steady insufflation pressures and the operating domain. Modern insufflation devices for laparoscopy have continuous pressure monitoring and automatic stop functions when pressures are met or exceeded. In neonates and young infants, flow rates are usually set to 1 liter per minute at the start of a procedure. The low flow rate can help ensure that a massive amount of air is not suddenly forced into the abdominal cavity, causing intra-abdominal pressures to exceed the preset limits. Flow rates are then titrated up as required to maintain the preset insufflation pressures. Flow rates generally increase up to 6 liters per minute in older children. However, set flow rates are not necessarily the rate of flow once instruments and the telescope are inserted into the trocars. If insufflation flow rates are not sufficient through the telescope trocar, a switch to one of the working ports is advised, as instruments are usually smaller in diameter compared to the telescope.

In infants and young children, many surgeons do not use trocars for the working instruments. With a thin abdominal wall, safe and efficient passage of an instrument does not require a trocar. However, the instrument access site may stretch or dilate as the case progresses, leading to a greater sustained loss of CO_2 through the incision(s). It is therefore necessary to increase flow rates to match or exceed ongoing gas losses, in order to maintain a working space.

Once the initial trocar has been successfully placed into the peritoneal cavity, the telescope is instrumental to safe and efficient placement of successive trocars into the peritoneal space. An angled telescope (e.g. 30-, 45-, or 70-degree bevel) allows for visualization of the peritoneal surface of the anterior abdominal. In addition, the course of the epigastric artery can be identified, so that it is not inadvertently injured during port placement. A superior epigastric artery injury can result in a very painful rectus sheath hematoma. An inferior epigastric artery injury will bleed vigorously, due to the lack of a posterior rectus sheath to tamponade the flow of blood.

Additional trocar placement methods depend upon the type of trocar being used. Reusable metal trocars are frequently use for pediatric laparoscopic procedures, particularly in infants and young children. These trocars come equipped with both sharp (cutting) and blunt tip obturators. A sharp-tip obturator requires a skin incision that approximates the diameter size of the trocar. The sharp obturator is then used to cut through the remaining musculofascial planes, as the trocar is advanced into position. The sharp tip of the obturator should be visualized at all times, to minimize the risk of inadvertent visceral injuries. A blunt-tip obturator requires a full-thickness incision to facilitate passage of the port. Under direct laparoscopic vision, an 11-blade scalpel is used to cut full-thickness through the skin and musculofascial planes. A fine dissecting instrument is then passed through the incision and a gentle spread opens up the incision to allow passage of the blunt obturator and trocar. Reusable metal trocars generally will require some type of suture to secure the trocar to the skin.

Additional radially expanding sheath trocars are placed using the same technique as for initial placement of a sheathed Veress needle technique, albeit with direct laparoscopic vision of the Veress needle. Once placed, the Veress needle is withdrawn and the trocar is advanced through the sheath. A gentle twisting motion of the trocar generally keeps the trocar moving straight through the sheath into the peritoneal cavity.

POSITIONING OF PORTS AROUND TARGET ANATOMY

In the vast majority of abdominal procedures, the umbilicus provides an excellent location for either a working instrument or the telescope. For procedures in the upper abdomen (e.g. cholecystectomy, splenectomy, fundoplication) or pelvis (oophorectomy, sigmoidectomy, rectal pullthrough), the telescope would remain in the umbilical port, with the target anatomy and the locations of the additional ports approximating a diamond configuration around the camera port. For procedures in the right and left paracolic gutters, the telescope is often moved out of the umbilicus, transitioning the umbilical port into a working port. For the vast majority of operations, the target anatomy forms the most distal point of a diamond shape, with the telescope located in the closest point of the diamond. The working instruments then form the right and left lateral points of the diamond. When more than three ports are required for the procedure, the location of additional ports is such that the working instrument through the additional port(s) will not interfere with the view from the telescope, or the work of additional instruments already inside the peritoneal cavity. The location of the ports for different procedures is beyond the scope of this introductory chapter, but will be detailed in the chapters specific to those procedures.

For some procedures, the telescope is moved laterally and instruments triangulated around the telescope. A common example is the laparoscopic Ladd's procedure. Placement of the telescope in the left lateral port allows excellent visualization of both the duodenum and the widening of the mesentery. However, in procedures with the telescope in a lateral port, the primary surgeon is then operating "off angle" from the usual triangulation with the telescope. This requires practice to become comfortable with the change in orientation on the screen. However, the early lack of comfort is frequently rewarded with improved visualization for the duration of the procedure.

When choosing the telescope and working ports, remember that this is not a static decision. If one struggles with a certain view, a change of the telescope to an alternate port can provide a different view of the anatomy. It is also advisable to remember that adding another port can also markedly improve retraction or visualization and improve efficiency for many operations.

RETROPERITONEAL PROCEDURES

A primary retroperitoneal dissection is begun with a 1 cm incision made between the twelfth rib and the iliac crest at the lateral margin of the quadratus lumborum. A trocar with a blunt-tip obturator in then advanced into the retroperitoneal space and a sweeping motion is begun with the trocar/obturator, to begin to dissect a space in the retroperitoneum. Gas insufflation is then begun, typically at a slightly higher insufflation pressure to tamponade the small bridge vessels in the retroperitoneum. Once additional ports are placed and the final working space is complete, the insufflation should be turned back down to usual levels for age, to prevent systemic absorption of excess CO_2.

Transperitoneal access is a very intuitive and logical approach to retroperitoneal structures, as laparoscopic access into the abdominal cavity is a common and comfortable procedure. Also, surgeons are more comfortable with anatomic variations and the relative positions of different organs when approaching transperitoneally. Access to retroperitoneal structures can be transmesocolic (through windows created in the colonic mesentery) or retromesocolic (via mobilization of the colon from surrounding attachments). Transmesocolic approaches are hampered by the small size of the mesenteric windows that can be safely created without injury to the mesenteric vasculature, and are more difficult in obese children and older adolescents with an abundant fatty mesentery. The key to transmesocolic approaches is to visualize the arborization of the mesenteric vessels, and to preserve all critical arcades of blood flow to the bowel. More commonly, the ascending or descending colon is mobilized medially, and/or the transverse colon is mobilized by entering into the lesser sac, to provide a wider dissection view for retroperitoneal structures.

CLOSURE OF LAPAROSCOPIC TROCAR SITES

The umbilical trocar site should always be closed, no matter the size of the trocar utilized or the age/size of the patient, as umbilical hernias are more likely to occur without fascial closure. However, closure of sites outside the umbilicus are influenced by the size of the fascial defect and the thickness of the abdominal wall and subcutaneous tissues. A general rule of thumb is to close the myofascial layer if it is visible through the skin incision, to prevent omental herniation through the trocar site. In older children and adolescents, myofascial closure of 5 mm ports is generally not required when located outside the umbilical ring. For 10–15 mm trocar sites, closure of a visible myofascial layer is recommended.

Myofascial closure is at risk of injury to the adjacent bowel or inclusion of the omentum within the closure. Protection of intra-abdominal contents from the sutures is greatly facilitated by use of a groove director. The groove director is placed into the incision along the peritoneal surface of the abdominal wall, then lifted away from the bowel. A 5/8 circle needle (Ethicon UR-6, Medtronic GU-46 or B. Braun FR26) allows for straight entry down to the groove director. Once it impacts the groove director, the hand is supinated to facilitate rotation within the groove, protecting underlying tissues from inadvertently being caught by the needle/suture. This process is repeated for figure-of-eight or simple interrupted sutures, as needed for the size of the fascial defect.

FURTHER READING

Murphy FJ, Mohee A, Khalil B, et al. Versatility of the circumumbilical incision in neonatal surgery. *Pediatr Surg Int* 2009; 25: 145–7.

Suri M, Langer JC. A comparison of circumumbilical and transverse abdominal incisions for neonatal abdominal surgery. *J Pediatr Surg* 2011; 46: 1076–80.

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* Charles McBurney (1845–1913), a New York surgeon who made significant contributions to the treatment of appendicitis.

* Hans Hermann Pfannenstiel (1862–1909), German gynecologist.

* Harrith M. Hasson (1931–2012), American gynecologist of Egyptian heritage working in Chicago.

* Janos Veres(s) (1903–1979), a Hungarian chest physician, designed this needle originally for initiating pneumothoraces in TB.

28

Fundoplication

DOUGLAS C. BARNHART AND ROBERT A. CINA

INTRODUCTION

Gastroesophageal reflux is a physiologic event which is more common in infants than in adults. This normal physiological occurrence is distinguished from pathological reflux (gastroesophageal reflux disease, GERD) by both the intensity (frequency and duration of reflux episodes) and the complications which arise as a consequence of the reflux. Despite a growing body of literature addressing diagnostic testing and long-term outcomes, the precise indications for Nissen fundoplication in specific patient populations remains controversial. Consequently, there is significant variability in the use of Nissen fundoplication between children's hospitals. This is particularly true with regard to infants and children with neurological impairment. Even considering these areas of controversy, Nissen fundoplication remains an important operation in the pediatric surgeon's armamentarium as it can definitively control GERD and thereby improve the quality of life and decrease the rate of hospitalization in selected patients.

HISTORY

In 1956, Rudolph Nissen of Basel, Switzerland, described the fundoplication procedure that he had been using for the previous 20 years to minimize postoperative reflux after resection of a peptic ulcer in the region of the cardia of the stomach. The technique did not involve division of the short gastric vessels, and the wrap extended for 4–6 cm on the esophagus. Subsequently, modifications have been made by Rossetti, Dor, Toupet, Donahue, and DeMeester. The first description of Nissen fundoplication using a laparoscopic approach in adults was by Dallemagne 1991. Subsequently Georgeson and Lobe separately reported initial experiences with laparoscopic Nissen fundoplication in pediatric patients in 1993.

PRINCIPLES AND JUSTIFICATION

Nissen fundoplication is indicated in infants and children who have complications of gastroesophageal reflux which are not adequately controlled with dietary modification and medical therapy. This principle can be difficult to operationalize because many symptoms attributed to gastroesophageal reflux can overlap with concomitant disease such as reactive airway disease, failure to thrive, feeding difficulties, irritability-related developmental delay, and cerebral palsy. Given these complexities, the decision about proceeding with fundoplication should be based on a detailed history with a particular emphasis on feeding issues and targeted diagnostic testing and should culminate with shared decision-making with the parents and other medical providers.

Clinical presentation

INFANT

Most infants with gastroesophageal reflux have only physiologic reflux and can be managed expectantly as the gastroesophageal reflux will decrease over the first year of life. Pathologic infantile gastroesophageal reflux is most often seen in children with neurological impairment. In these children the most common problems related to gastroesophageal reflux are failure to thrive and pulmonary complications such as aspiration pneumonia. Nissen fundoplication is often performed with concomitant gastrostomy tube placement. Pathologic gastroesophageal reflux which cannot be successfully managed with feeding manipulations and medical therapy is uncommon in neurologically normal infants. The most common scenario in these children is failure to thrive.

OLDER CHILD

Older children present with gastroesophageal disease with more typical esophageal symptoms. This often includes heartburn and waterbrash. Regurgitation with activity and positional changes occurs. Dysphagia can occur due to either esophagitis or a stricture. Exacerbation of reactive airway disease may be a complication of gastroesophageal reflux but attribution is challenging in individual patients.

NEUROLOGICAL IMPAIRMENT

Children with neurological impairment have frequent foregut symptoms which can include retching after feedings, poor tolerance of gastric feedings, and frequent emesis. Failure to thrive is also common in these patients. These symptoms may be due to gastroesophageal reflux but can also be due to other causes such as delayed gastric emptying, esophageal dysmotility, and centrally mediated hypertonicity. Decision-making regarding fundoplication is particularly challenging in these patients due to the multiple causes of these symptoms and the increased risk of postoperative recurrent gastroesophageal reflux in these children.

ANATOMIC ANOMALY

Gastroesophageal reflux is very common in children who have undergone repair of esophageal atresia. Decision-making regarding fundoplication is complicated by the decreased esophageal motility in these patients. Pathologic reflux is also more common in infants with congenital diaphragmatic hernia, anterior abdominal wall defects, and malrotation.

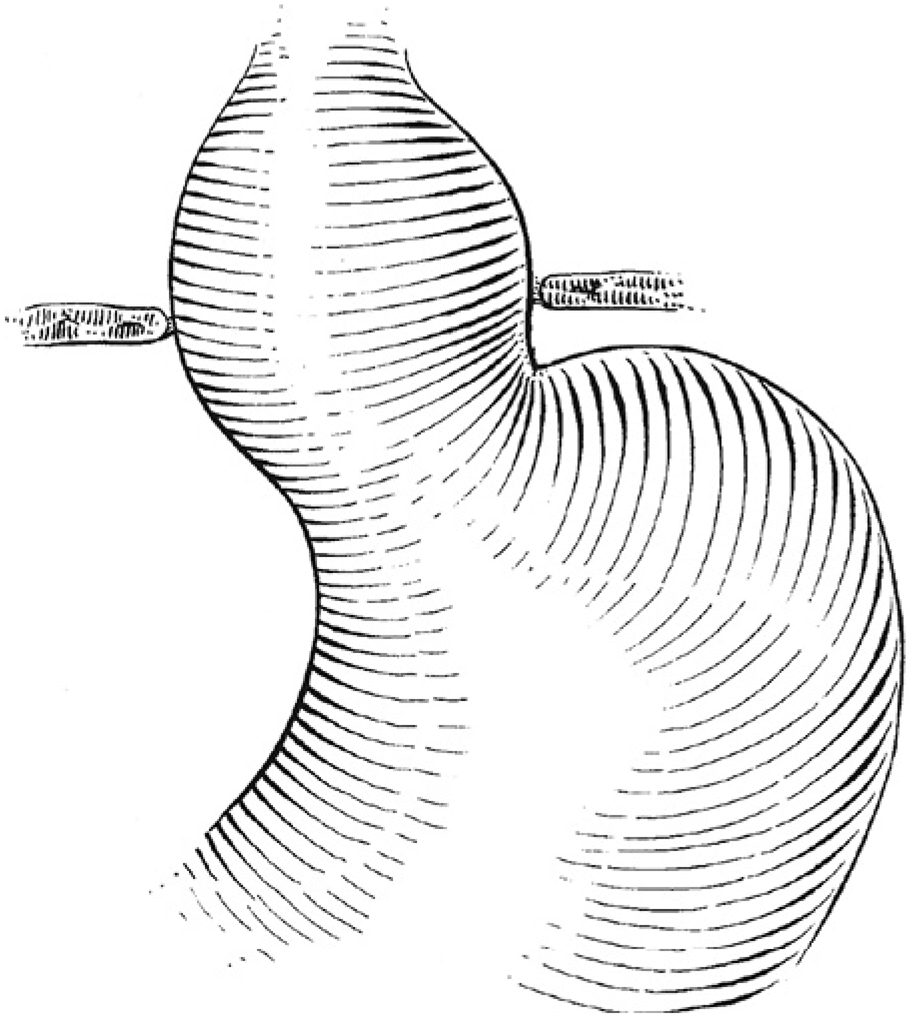
OTHER UNUSUAL PRESENTATIONS

Other presentations include rumination, Sandifer’s syndrome (spastic torticollis and spasmodic, rigid opisthotonic posturing), irritability, and hyperactivity.

Pathologic anatomy

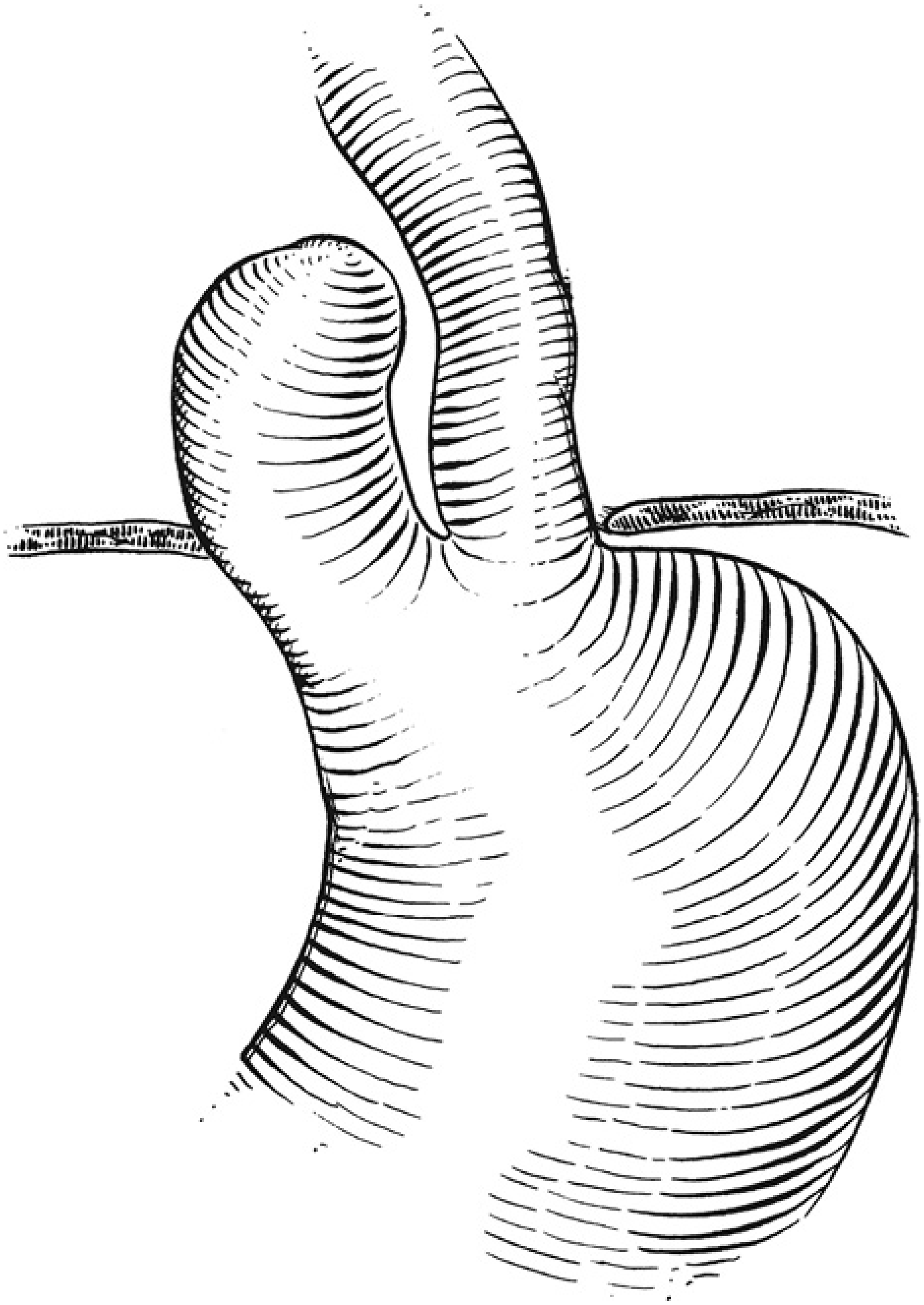
The majority of children with pathologic gastroesophageal reflux have normal foregut anatomy. Hiatal hernias are more common in children with a history of esophageal atresia and congenital diaphragmatic hernia. Hiatal hernias can occur in otherwise normal children with GERD but are uncommon. Two types of hiatus hernia are recognized:

- *Sliding hiatus hernia* is characterized by ascent of the cardia into the mediastinum (**Figure 28.1**).



28.1

- In *paraesophageal or rolling hernia*, the gastroesophageal junction remains in the abdomen while part of the gastric fundus prolapses through the esophageal hiatus into the mediastinum (**Figure 28.2**).



28.2

The sliding hernia is often associated with reflux, while gastric stasis in the paraesophageal hernia predisposes to peptic ulceration, perforation, and hemorrhage.

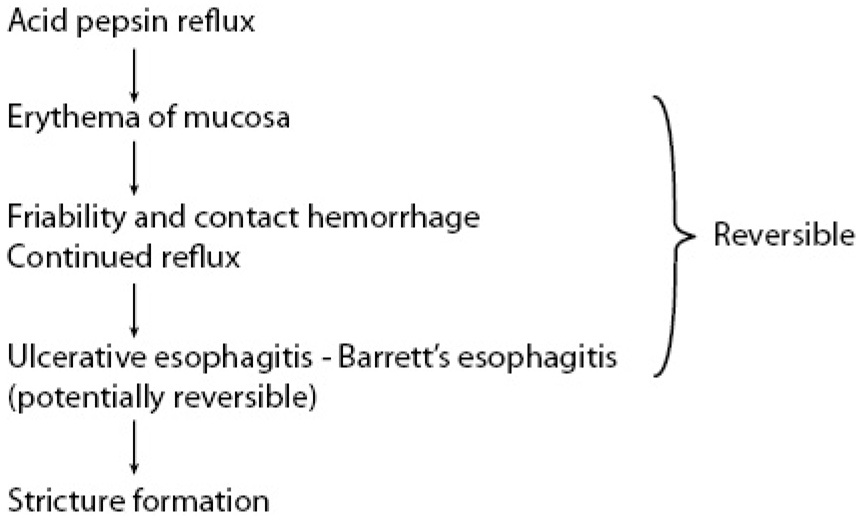
NORMAL MECHANISMS PREVENTING REFLUX

Physiologic control of reflux depends on the following factors:

- Anatomic
  - Length and pressure of the lower esophageal sphincter
  - Intra-abdominal segment of the esophagus
  - Gastroesophageal angle (angle of His)
  - Lower esophageal mucosal rosette
  - Phrenoesophageal membrane
  - Diaphragmatic hiatal pinchcock effect
- Physiologic
  - Coordinated effective peristaltic clearance of the distal esophagus
  - Normal gastric emptying

PATHOPHYSIOLOGY OF REFLUX

The squamous epithelium of the esophagus is unable to resist the irritant effect of gastric juices. The acid pepsin causes a chemical inflammation with erythema of the mucosa. With continued reflux, the mucosa becomes friable and bleeds easily on contact. Later, frank ulceration develops, which, with repeated attempts at repair and relapse, eventually leads to stricture formation. This process is summarized in **Figure 28.3**.


Acid pepsin reflux
Erythema of mucosa
Friability and contact hemorrhage
Continued reflux
Ulcerative esophagitis - Barrett's esophagitis
(potentially reversible)
Stricture formation
Reversible

28.3 Pathophysiology of reflux.

INDICATIONS FOR ANTIREFLUX SURGERY

Antireflux procedures are indicated when there are ongoing complications of gastroesophageal reflux despite maximal medical therapy. This is clearly the case when children have esophagitis or an esophageal stricture despite effective acid suppression. These diagnoses should be objectively confirmed as symptoms are notoriously non-specific. Other indications for antireflux surgery in children may include failure to thrive or pulmonary exacerbations. These are more difficult to specifically determine. In the absence of ongoing esophagitis, a thorough diagnostic evaluation for other causes of failure to thrive should be completed. While some patients with asthma may have exacerbations triggered by gastroesophageal reflux (via other vagal mediated neurologic) or microaspiration, it is difficult to attribute these exacerbations confidently to gastroesophageal reflux.

The most common setting for antireflux procedures in infancy is with concomitant gastrostomy tube placement. There is wide variation between centers with regard to the prevalence of antireflux procedures at the time of gastrostomy tube placements. This reflects the lack of diagnostic precision in defining pathologic reflux in infants and challenges in attribution of pulmonary events to reflux. A practical approach is to compare the occurrence of these events during nasogastric and nasojejunal feedings. Fundoplication should be considered for infants who have increased frequency of vomiting, feeding intolerance, or pulmonary events during gastric feedings.

Repeated episodes of aspiration pneumonia in children with neurological impairment can be an indication for antireflux procedures as well. In these children a quantitatively normal degree of gastroesophageal reflux may still be problematic if the child has failure to adequately protect the airway. Even infrequent episodes of gastroesophageal reflux may result in aspiration pneumonia. It is important to acknowledge that antireflux procedures do not eliminate the risk of aspiration pneumonias as these may occur secondary to oral secretions too.

PREOPERATIVE INVESTIGATIONS

The use of diagnostic studies prior to antireflux surgery varies with age and the clinical presentation. Not all studies need to be completed in all patients.

Surgeons should consider the specific clinical scenario and the issues which need clarification in determining which studies are appropriate.

Upper gastrointestinal contrast study

All patients should undergo a contrast study of the foregut prior to fundoplication. This allows identification of obstructive lesions of the esophagus, stomach, or duodenum which could cause symptoms similar to gastroesophageal reflux. It also allows clarification of the esophageal anatomy and provides a qualitative assessment of clearance. Hiatal hernias can typically be demonstrated. The upper gastrointestinal (GI) contrast study is not reliable for quantification of gastroesophageal reflux, however. The degree of reflux seen is dependent upon the volume of contrast administered, period of observation, and agitation of the child during the examination.

Endoscopy and biopsy

Endoscopy can provide important confirmation that symptoms of dysphagia and chest pain are related to esophageal mucosal disease. Endoscopic visualization allows determination of the extent of involvement and severity by standardized grading scores. Four grades of esophagitis are recognized at endoscopy:

- Grade I, erythema of mucosa
- Grade II, friability of mucosa
- Grade III, ulcerative esophagitis
- Grade IV, stricture

Endoscopic biopsies should routinely be performed during endoscopy for foregut symptoms. Importantly, these biopsies will allow distinction between reflux esophagitis and eosinophilic esophagitis. Eosinophilic esophagitis should not be primarily treated with antireflux surgery. Histology of the biopsy will provide pathologic grading of reflux esophagitis.

Esophageal pH monitoring

Continuous 24-hour monitoring of esophageal pH can be done in the distal esophagus (single channel) or in multiple locations of the esophagus (multiple channel). This is the most precise method to quantify the extent of acid reflux with clearly established norms. A pH of less than 4 is regarded as significant. Multiple parameters may be measured and assessed individually as well as being summarized in various scoring systems. The simplest score is the reflux index (RI), which is percentage of time with pH <4, with the upper limit of normal being 3% and 4–7% being borderline. During the 24-hour recording, the following parameters should be examined:

- Number of episodes during which the pH falls below 4
- Duration of each reflux episode
- Number of episodes lasting more than 5 minutes
- Total duration of reflux, expressed as a percentage of recording time

Combined multiple intraluminal impedance and pH monitoring

In addition to measurement of pH, multiple intraluminal impedance (MII) monitoring allows the detection of non-acid reflux. Non-acid reflux may also cause esophagitis and esophageal symptoms, and this technique allows quantitative assessment and comparison to norms. Due to the number and fast response of the detectors, this monitoring allows the timing and anatomic location of reflux to be accurately detected. This is particularly advantageous in assessing symptom correlation with reflux episodes. This can be useful in correlating whether pulmonary and airway symptoms are temporally related to gastroesophageal reflux. pH monitoring alone does not have a quick enough response to allow this assessment.

Esophageal manometry

Pressure recordings are made with continuously perfused, open-tipped catheters or solid-state pressure transducers. This allows assessment of resting pressures within the esophagus and assessment of esophageal motility. These findings are not specific or sensitive enough to diagnose pathologic gastroesophageal reflux. Manometry is useful in diagnosing alternative causes of esophageal symptoms such as motility disorders.

Scintiscanning

Technetium (^{99}Tc) sulfur colloid scans may be useful in documenting pulmonary aspiration. Standards for use of this test for gastroesophageal reflux are, however, poorly defined.

MEDICAL MANAGEMENT

Non-operative treatment of GERD should include lifestyle and dietary modifications as well as pharmacologic treatment. The most effective non-pharmacologic treatment in infants is alteration of the volume and frequency of feeding. Thickening of feedings does not decrease the frequency of measurable reflux episodes although it may reassure parents by decreasing the number of visible episodes of regurgitation. The only position shown to decrease esophageal acid exposure is prone but this should not be recommended due to the increased risk of sudden infant death syndrome (SIDS). Lifestyle modification in older children can include weight loss, avoidance of large meal volume, and prone or left-sided sleeping.

Acid suppression is the mainstay of modification. Two classes of drugs, histamine-2 receptor antagonists and proton pump inhibitors, are commonly used for acid suppression. Each of these classes and specific agents has side effect profiles but they are generally well tolerated. Evidence suggests that acid suppression may be associated with an increased risk of community-acquired pneumonia. Currently available pro-kinetic agents have low efficacy and numerous side effects and therefore are not recommended for general use in the treatment of GERD.

Medical treatment and diagnostic consensus guidelines have been jointly defined by the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN).

CHOICE OF OPERATIVE TECHNIQUE

Multiple factors influence the choice of operative approach including surgeon experience, available resources, and individual patient factors. Additional factors which should be considered are resource utilization (operative time and hospital length of stay), postoperative pain, speed of recovery, and durability. Three randomized controlled trials (RCTs) have compared open and laparoscopic Nissen fundoplication. One of these showed a higher rate of recurrent reflux requiring reoperation in the open approach and another trended in this direction but did not reach significance. Large administrative database analyses have demonstrated the laparoscopic approach to be associated with decreased resource utilization and to have a favorable adverse event rate. Several small RCTs have not shown any consistent differences between partial and complete fundoplications.

Two technical aspects of laparoscopic Nissen fundoplication have been examined in separate RCTs. The first showed that minimal esophageal dissection at the hiatus decreased the rate of transhiatal migration of the fundoplication and the need for reoperation. The second showed that suture fixation of the esophagus to the crura did not decrease transhiatal migration.

ANESTHESIA

General endotracheal anesthesia is administered, with the patient supine. The use of perioperative prophylactic antibiotics is controversial in fundoplication without gastrostomy as this is a clean operation. If the surgeon chooses to use prophylaxis, this should typically be limited to cefazolin. Epidural analgesia should be considered for open fundoplication as it may control postoperative pain and decrease respiratory complications.

LAPAROSCOPIC NISSEN FUNDOPLICATION

Principles and justification

Laparoscopic Nissen fundoplication offers several advantages over the open technique, is both safe and effective, and can be broadly applied to most pediatric patients. The major contraindication to this technique is the child's inability to tolerate a pneumoperitoneum due to underlying respiratory or cardiovascular compromise. While previous abdominal surgery may make successful execution of this technique more challenging, adhesive disease confined to the lower abdomen is rarely problematic, and left upper quadrant adhesions, if present, are typically amenable to laparoscopic adhesiolysis. The presence of a previous gastrostomy makes the operation slightly more challenging, although moving the camera port medial to the gastrostomy allows the procedure to proceed with minimal additional difficulty. In rare instances, the gastrostomy can be taken down from the abdominal wall, temporarily closed with intracorporeal sutures, and a new tube placed through the old gastrostomy at the end of the procedure using the standard laparoscopic U-stitch technique.

Preoperative preparation and anesthesia

Air trapped within the GI tract can make visualization of the pertinent anatomy tedious, particularly when the small bowel and colon are distended. In select patients, nasogastric decompression for 12 hours preoperatively helps to minimize this. In addition, prompt removal of air introduced into the GI tract during intubation is extremely valuable.

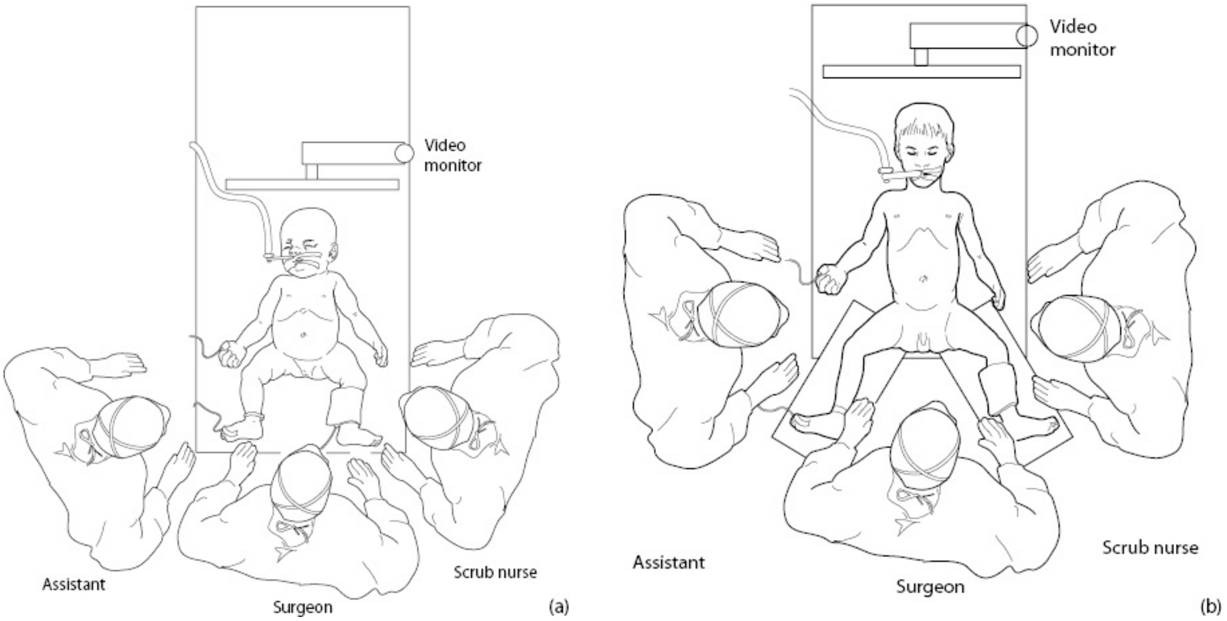
ANESTHESIA

Constant communication with the anesthetic team is critical, particularly in patients with chronic respiratory failure and complex congenital heart disease. The judicious use of pneumoperitoneum and constant awareness of the sequelae of pneumoperitoneum on neonatal physiology are necessary. Typically, we try to limit the pneumoperitoneum pressure to 8–12 cm water in neonates, and use a maximum pressure of 15 cm water in older children.

Placement of a bougie can facilitate the identification of the esophageal wall, and allow for the safe execution of the retroesophageal dissection. Following positioning of the patient, the stomach is decompressed with an oral gastric tube, and an appropriately sized bougie (sized to the diameter of the patient's thumb) can be placed into the stomach.

PATIENT AND TEAM POSITIONING

Infants are placed in the frog-leg position at the foot of the bed, all pressure points are padded, and we ensure that they are adequately secured to the bed by tilting the bed in reverse Trendelenburg (15–30 degrees). Careful attention should be made to route all cords so as to not interfere with the securing of rail-mounted retractors to the bed. The primary monitor is placed above the head of the bed, just below eye-level to facilitate surgeon ergonomics (**Figure 28.4a**).



28.4a,b

In older children, the legs are placed in pneumatic stirrups such that the thighs are in line and parallel to the bed, with the legs bent at the knee (**Figure 28.4b**). Care must be taken to ensure the avoidance of common peroneal nerve compression.

LAPAROSCOPIC INSTRUMENTATION

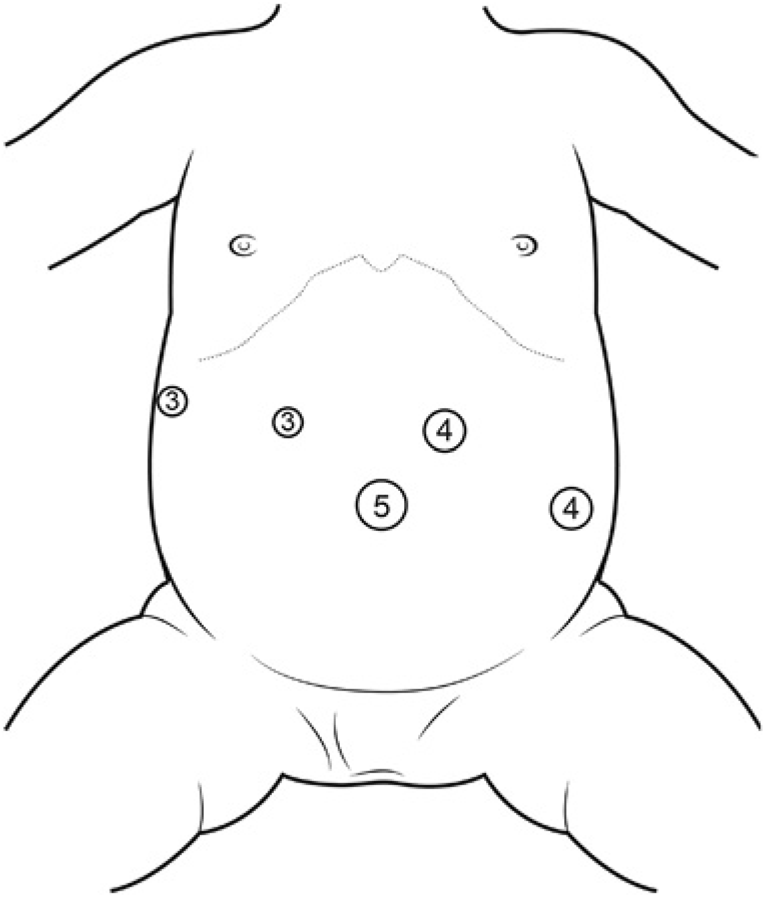
The following instruments are required:

- 5 mm 30-degree telescope
- 3 mm or 5 mm trocar × 4
- 5 mm radially expanding disposable trocar
- Rail-mounted flexible "snake" liver retractor
- Atraumatic fenestrated intestinal graspers
- Atraumatic fenestrated duckbill graspers
- Needle holder
- Hook electrocautery, advanced bipolar or ultrasonic sealer
- Suture scissors
- 2-0 Neurolon or Silk on a SKI (PB-1) needle
- Enteral Access Introducer Kit guidewire, needle, sequential dilators (8-20 Fr)
- Low profile gastrostomy

Operation

PORT PLACEMENT

Five ports are used to perform the fundoplication (**Figure 28.5**). Attention is first turned to the umbilicus, where the skin is incised vertically in the base of the umbilicus after infiltrating with local anesthetic. The underlying fascia is grasped with mosquito snaps and also incised. A mosquito snap is then placed through this fascial defect into the peritoneal cavity, ensuring free entry into the peritoneum. A Veress needle with a radially expanding introducer sheath is then placed through this fascia defect using modified open technique. Following removal of the needle, a 5 mm trocar is placed through this. A pneumoperitoneum is established at a pressure of 8–12 cm of water with carbon dioxide insufflation.

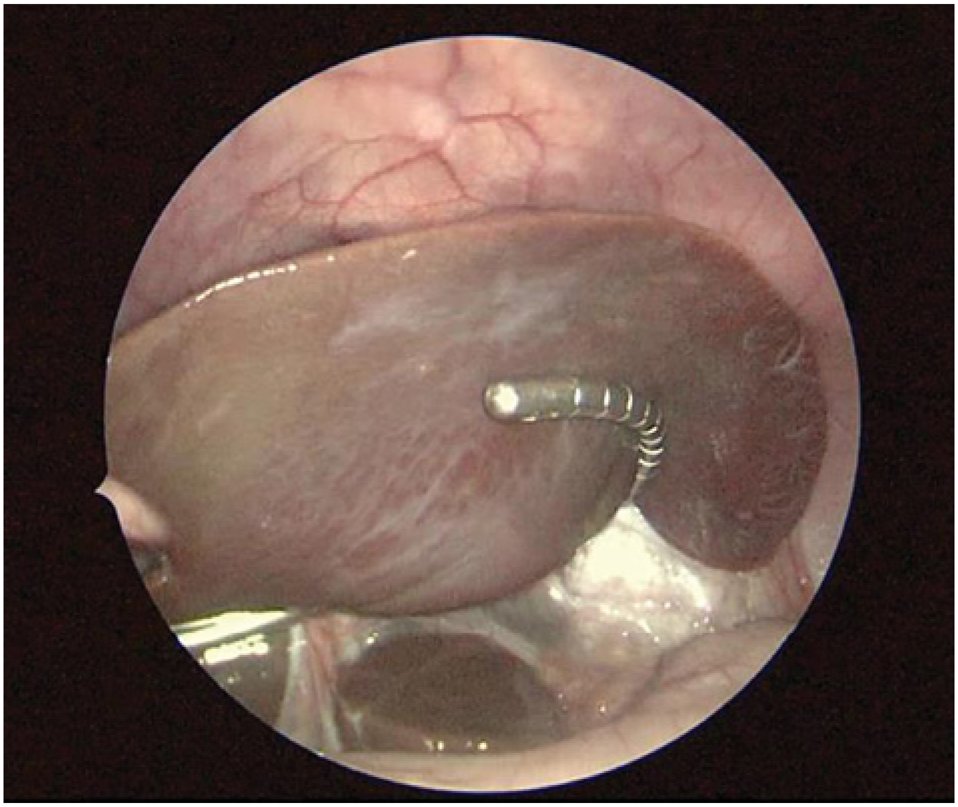


28.5

Four additional ports are placed (3 mm or 5 mm depending on the size of the patient). Two ports are placed on the patient's left. The first of these is placed approximately halfway between the umbilicus and the costal margin just to the left of midline. This will serve as the main camera port during the operation and ultimately will serve as the site of the gastrostomy. An additional 4 mm port is placed in the anterior axillary line between the costal margin and the anterior superior iliac spine, and this will serve as the primary port for the cautery, blunt grasper, and needle driver. Two additonal ports are also placed, inferior to the costal margin medial to the right mid-clavicular line, with the second quite lateral in the right subcostal area to accommodate the liver retractor. The mid-clavicular line port serves primarily as the surgeon's left-hand instrument.

LIVER RETRACTION

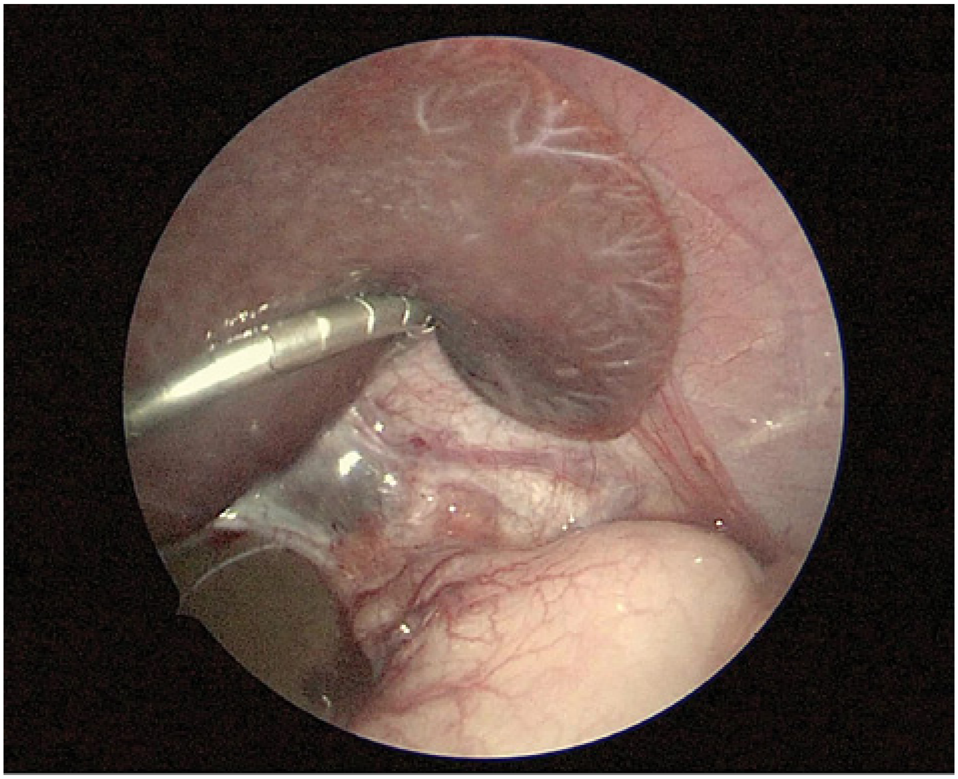
Multiple techniques may be used to retract the liver. An appropriately sized Nathanson liver retractor is then used to lift the liver up and expose the esophageal hiatus and is secured with a table-mounted flexible retractor. Alternatively, a flexible or fan retractor can be introduced via the right upper quadrant. If none of these retractors is available, a locking grasper can be placed through an epigastric port and used to hold the left lateral segment anteriorly by grasping the right crus superiorly. Once the esophageal hiatus is adequately exposed, the camera is then placed in the left upper quadrant port (**Figure 28.6**).



28.6

EXPOSURE OF THE CARDIOESOPHAGEAL JUNCTION

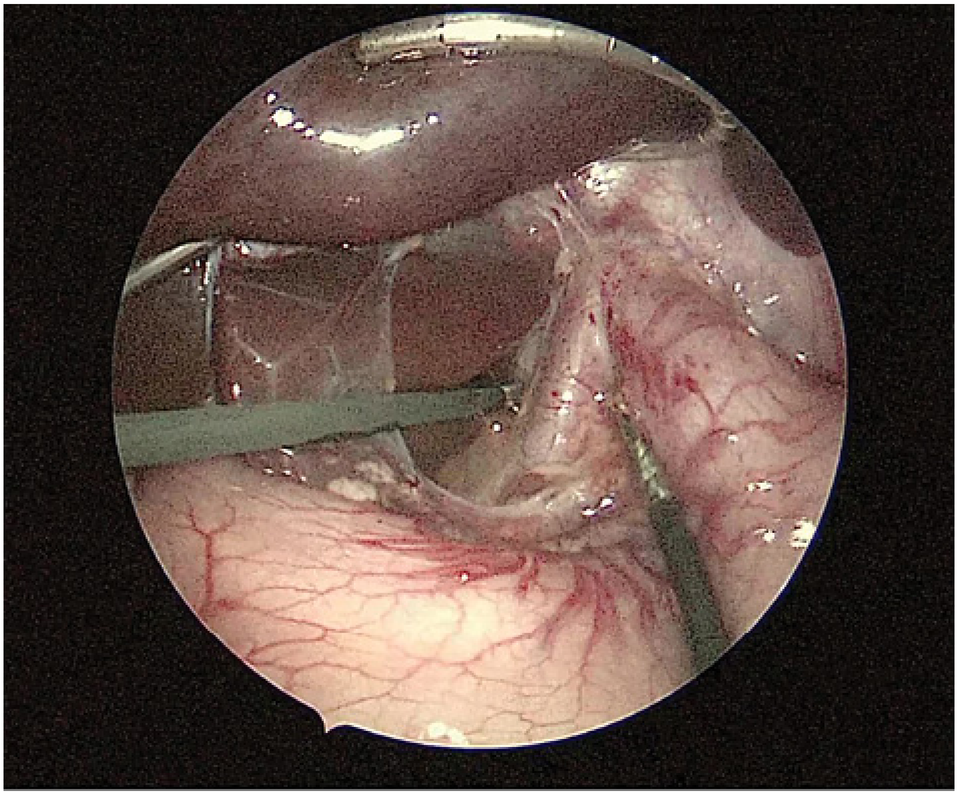
Although some teams prefer to start with the intra-abdominal portion of the operation, the authors begin with exposure of the cardioesophageal junction. The stomach is grasped with an atraumatic grasper by the assistant and retracted to the patient's left. Using a blunt grasper, the pars flaccida is grasped and the hook electrocautery is used to incise the gastrohepatic ligament. Care must be taken during this step to determine the presence of a replaced or accessory left hepatic artery, which is present in approximately 15% of cases and arises from the left gastric artery (**Figure 28.7**)]. Preservation of this is optimal, but division is usually safe when necessary. Once the gastrohepatic ligament is divided up to the phrenoesophageal junction, the gastroesophageal junction is grasped and the esophagus can be rolled to the patient's left, exposing the right crus.



28.7

CRURAL EXPOSURE

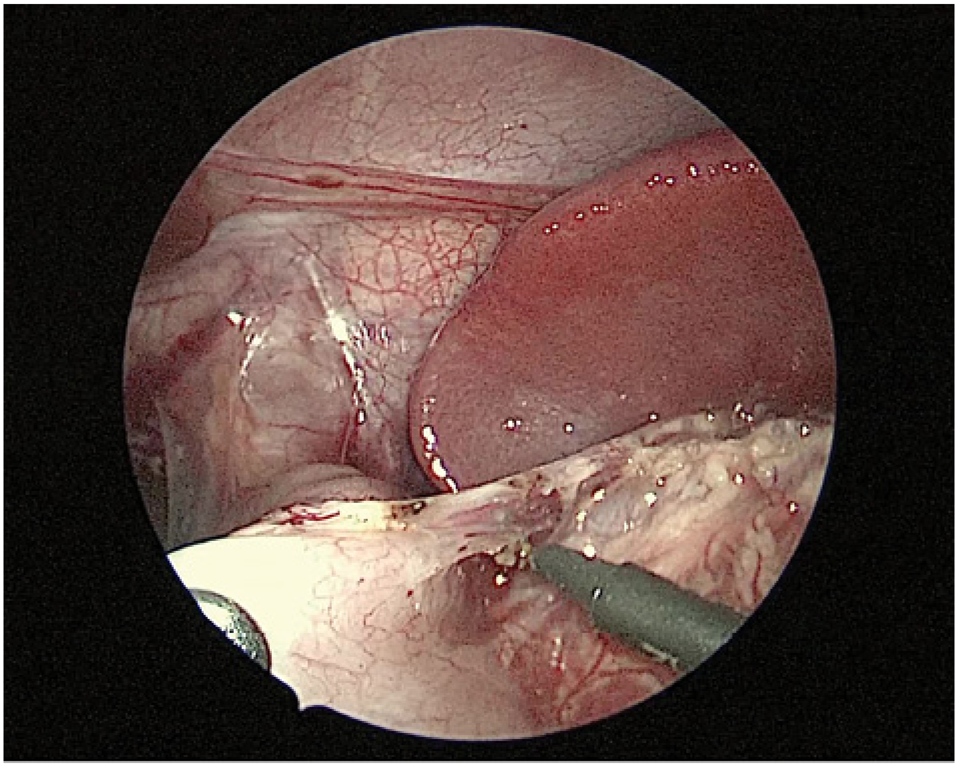
The peritoneum at the junction between the right crus and the esophagus is scored with the electrocautery to allow for a blunt retroesophageal dissection (**Figure 28.8**). It is helpful to ensure that the preoperatively placed bougie is still in position before proceeding with this dissection. A minimal dissection technique is used, to achieve only what is necessary to safely visualize the pertinent anatomy. It is imperative that the retroesophageal window is made above the left gastric artery, and that the right and left crus can be identified. The fascia overlying the crura should be left intact, and the crura should not be divided. A key technical point during this step is that the dissection be directed transversely, as an oblique dissection can lead to injury of the left crura or left pleural. Surgeons vary in terms of routine placement of crural sutures. We do not advocate routine cruraplasty, reserving this maneuver for the presence of a hiatal hernia. When a cruraplasty is necessary, placement of theses sutures can be facilitated by pulling the bougie in the proximal esophagus to allow for better visualization of this area. This is performed with either 2/0 braided Nylon (Nurolon•) or 2/0 silk on a SKI or BB tapered needle.



28.8

DIVISION OF THE SHORT GASTRIC VESSELS

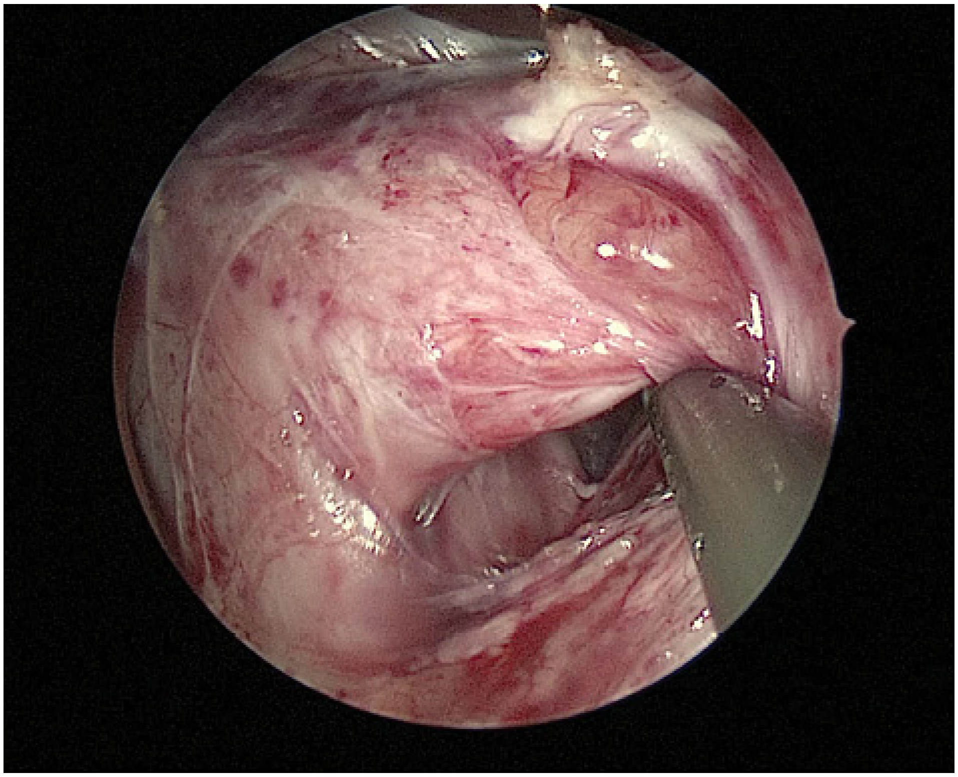
Attention is then turned to the left upper quadrant, and the stomach is grasped along the greater curvature and retracted to the right, exposing the gastrosplenic ligament. The cranial portion of this, along with the short gastric vessels, is divided using electrocautery (in infants and toddlers), or with an advanced energy device (LigaSure™) in older children and teenagers (**Figure 28.9**). Care must be taken to avoid thermal injury to the stomach, particularly as the division proceeds cephalad. It is also important to note that the gastrosplenic ligament splays to form anterior and posterior leaves at this location, and that both need to be divided. Once complete, the left crus is easily identified.



28.9

CREATION OF POSTERIOR ESOPHAGEAL WINDOWS

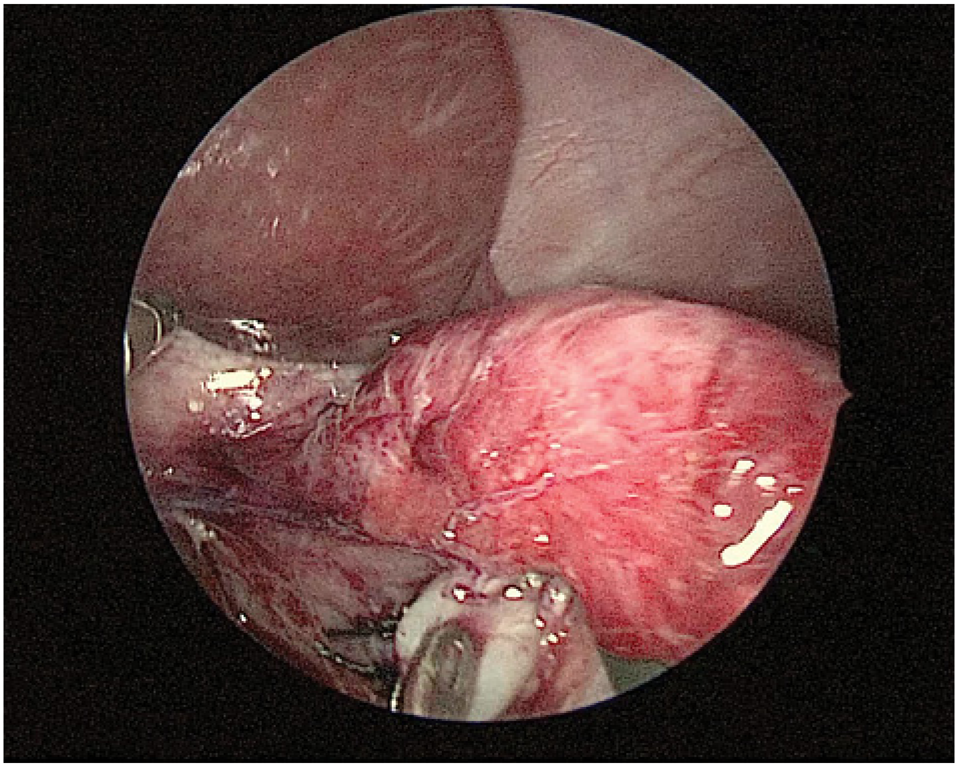
Attention is again turned to the previous retroesophageal dissection, and the posterior esophageal window is completed using blunt dissection with two non-traumatic graspers. Once complete, it should be possible to visualize the spleen or the fundus of the stomach through the retroesophageal window (**Figure 28.10**).



28.10

FUNDAL MOBILIZATION

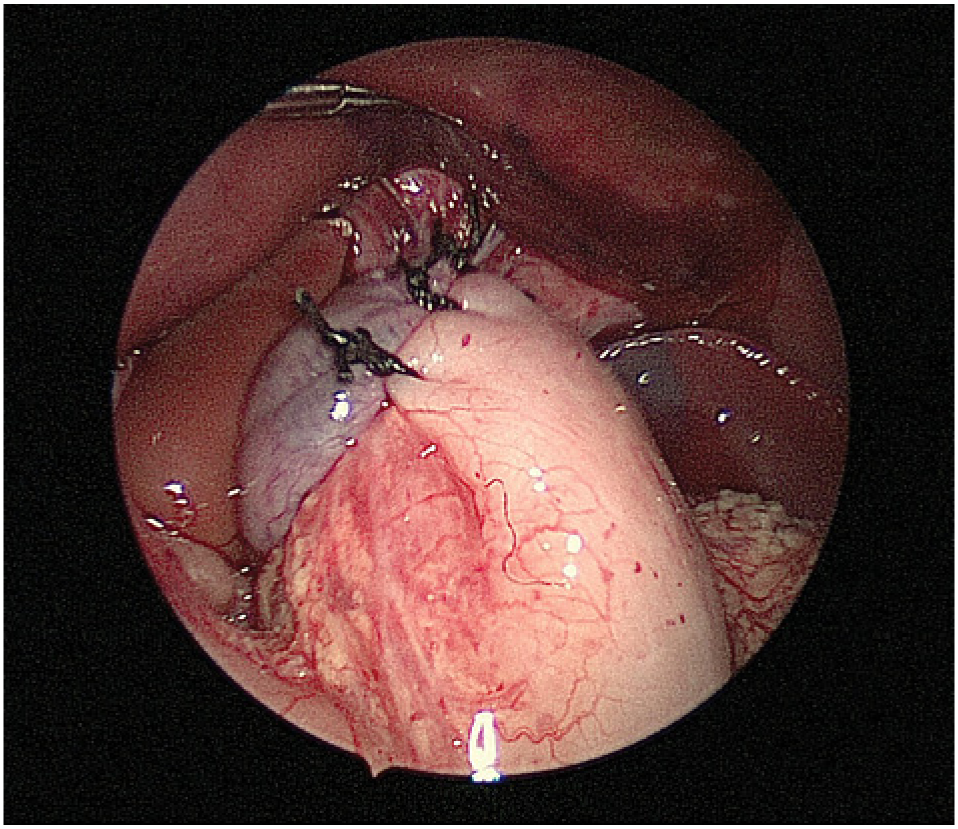
The bougie is temporarily brought back into the esophagus, and the gastric fundus grasped and brought through the retroesophageal window. Once brought through, it should lie with minimal tension. A “shoe-shine” maneuver should be performed to ensure that the fundus is not twisted as it passes through the retroesophageal window (**Figure 28.11**).



28.11

CREATION OF THE WRAP

Three to five sutures are then utilized to create an anterior 1–2.5 cm fundoplication with 2/0 silk or braided nylon on a SKI needle. The length of the fundoplication varies with age and is the same as described earlier for the open technique. While a variety of techniques can be employed to tie these, we find intracorporeal knot tying to be fast and efficient. The first suture (typically 12 cm in length) is placed in the anterior wall of the stomach, incorporating the anterior wall of the esophagus, and the leading edge of the fundus. It is helpful to have the assistant grasp the gastroesophageal junction and retract this down as the first suture is tied. Once secured, one or two additional sutures (10 cm in length) can be placed cephalad to this first suture (which also incorporate esophagus), with two additional sutures placed caudal to this (**Figure 28.12**). Following placement of these sutures, the bougie is slowly removed by the anesthetic team to ensure that it was not incorporated in the closure. The wrap should be floppy without tension on the fundus to minimize postoperative dysphagia.



28.12

INSERTION OF LAPAROSCOPIC-ASSISTED GASTROSTOMY

At this point, the liver retractor is removed under laparoscopic visualization and the camera is placed in the umbilical port site. A 3 mm blunt grasper is placed through the 4 mm left upper quadrant port and used to manipulate the stomach. The stomach is grasped along the greater curvature, and the site for the gastrostomy chosen. Two 0/PDS sutures are then placed through the abdominal wall, through the stomach, and back out through the abdominal wall on each side of the grasper. The grasper is then removed from the abdomen, and a needle placed through the incision site and visualized touching the gastric wall.

Once the anesthetic team insufflates the stomach with air, the stomach is cannulated with a needle and guide wire. An 8 Fr dilator is then placed through the gastrostomy using wire guidance. Upon withdrawal of the dilator, the abdominal wall thickness should be measured, and an appropriate tube chosen. The gastrostomy tract is then dilated using the 12 and 16 Fr dilators. Next, the gastrostomy tube is placed into the stomach with the smallest dilator passed through its central lumen, using wire guidance. The wire and dilator are removed, and tubing was attached to the gastrostomy. The stomach is insufflated with air through this tube using a catheter-tipped syringe to ensure intraluminal placement. Finally, the sutures are secured over the tube.

CLOSURE

The abdomen should then be desufflated and the fascia closed with absorbable sutures. The skin can be closed with Dermabond® or other skin glue.

Postoperative care

Routine gastric decompression is not required in routine cases. If a gastrostomy tube is inserted, the nasogastric tube is removed at the end of the operation and the gastrostomy tube left on drainage. Oral or gastrostomy feeds are started on the first postoperative day. Patients should remain on soft solid diet for the first 2 weeks after fundoplication, gradually increasing the consistency of the food.

Acid suppressive therapy is continued postoperatively. The dose of this is cut in half at 2 weeks postoperative, and discontinued at the 1 month postoperative visit to prevent gastric acid hypersection from sudden cessation.

Complications

An immediate postoperative complication is dysphagia, which, if sufficiently symptomatic, can be relieved with an early flexible endoscopy or, if not responding, by performing single balloon dilatation. Retching postoperatively can occur, particularly in children with neurological impairment. Complication management is similar to that described in "Open fundoplication: Complications."

Outcome

Laparoscopic fundoplication generally has favorable outcomes. Unfortunately, neurologically impaired patients have a higher incidence of wrap migration, disruption, and failure to correct reflux than their neurologically normal counterparts.

OPEN FUNDOPLICATION

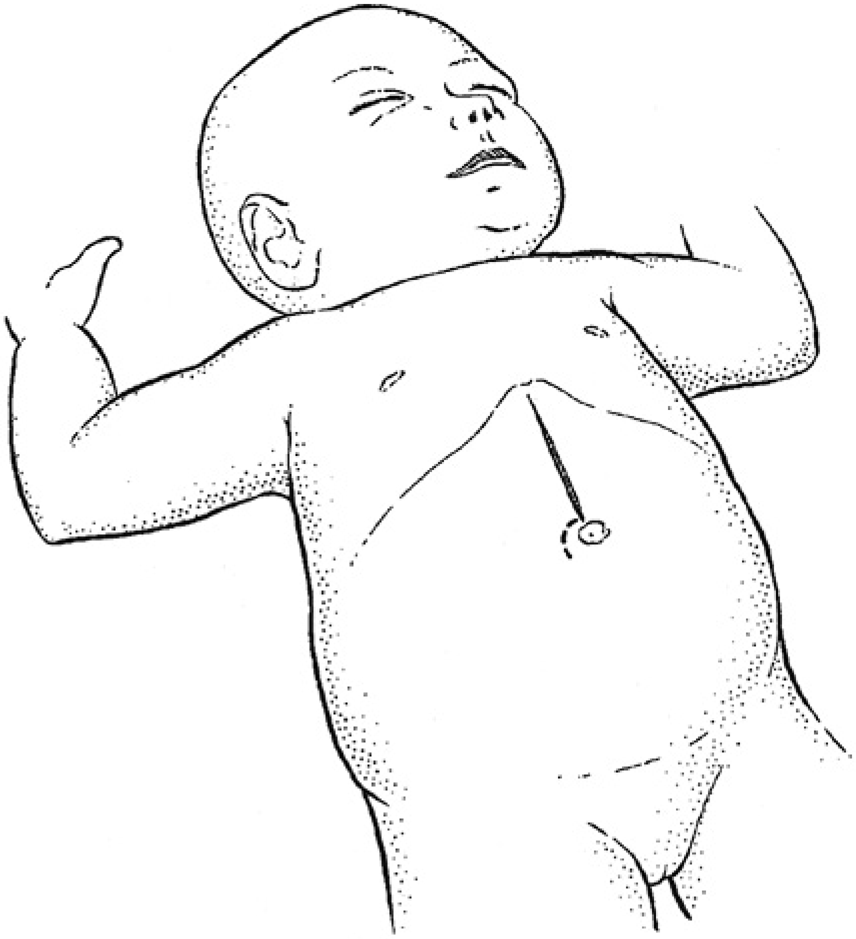
Anesthesia

Placement of a large-caliber bougie can facilitate identification of the esophagus by palpation, aid in the identification of a partial thickness injury, and prevent the cruroplasty and fundoplication from being overly tight. This should be selected to distend but not dilate the esophagus. A useful guide is the diameter of the patient's thumb. Some surgeons place only a nasogastric tube.

Operation

INCISION

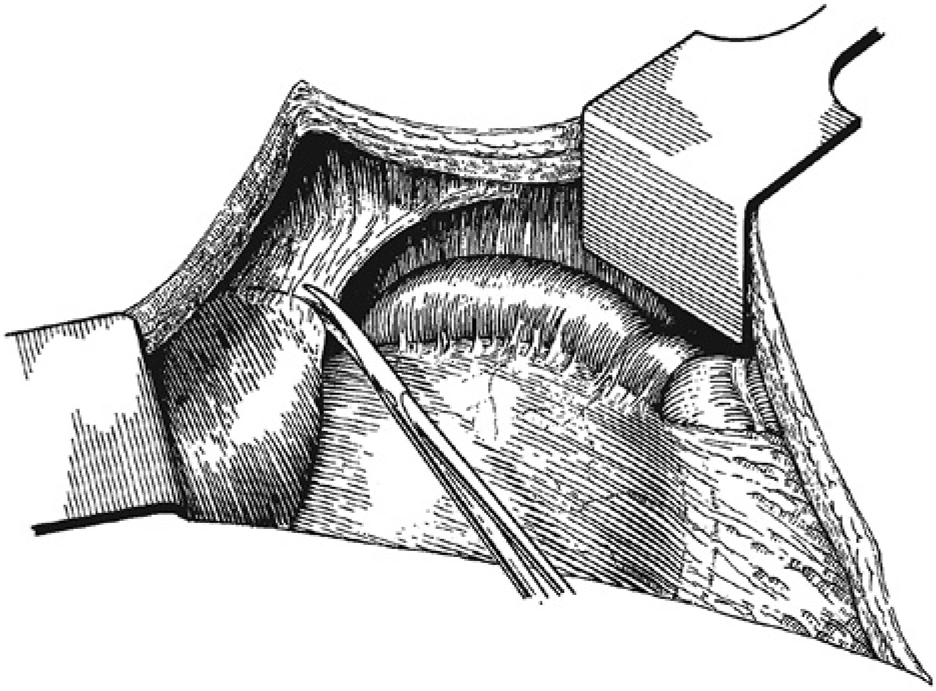
Excellent exposure can be obtained using either a supraumbilical midline incision (**Figure 28.13**) or a left subcostal incision. In infants who have had a previous supraumbilical transverse laparotomy, this approach can be used if fixed traction is placed. As many surgeons use open fundoplication mainly as a secondary procedure, the choice of incision is often based on the prior scar.



28.13

EXPOSURE

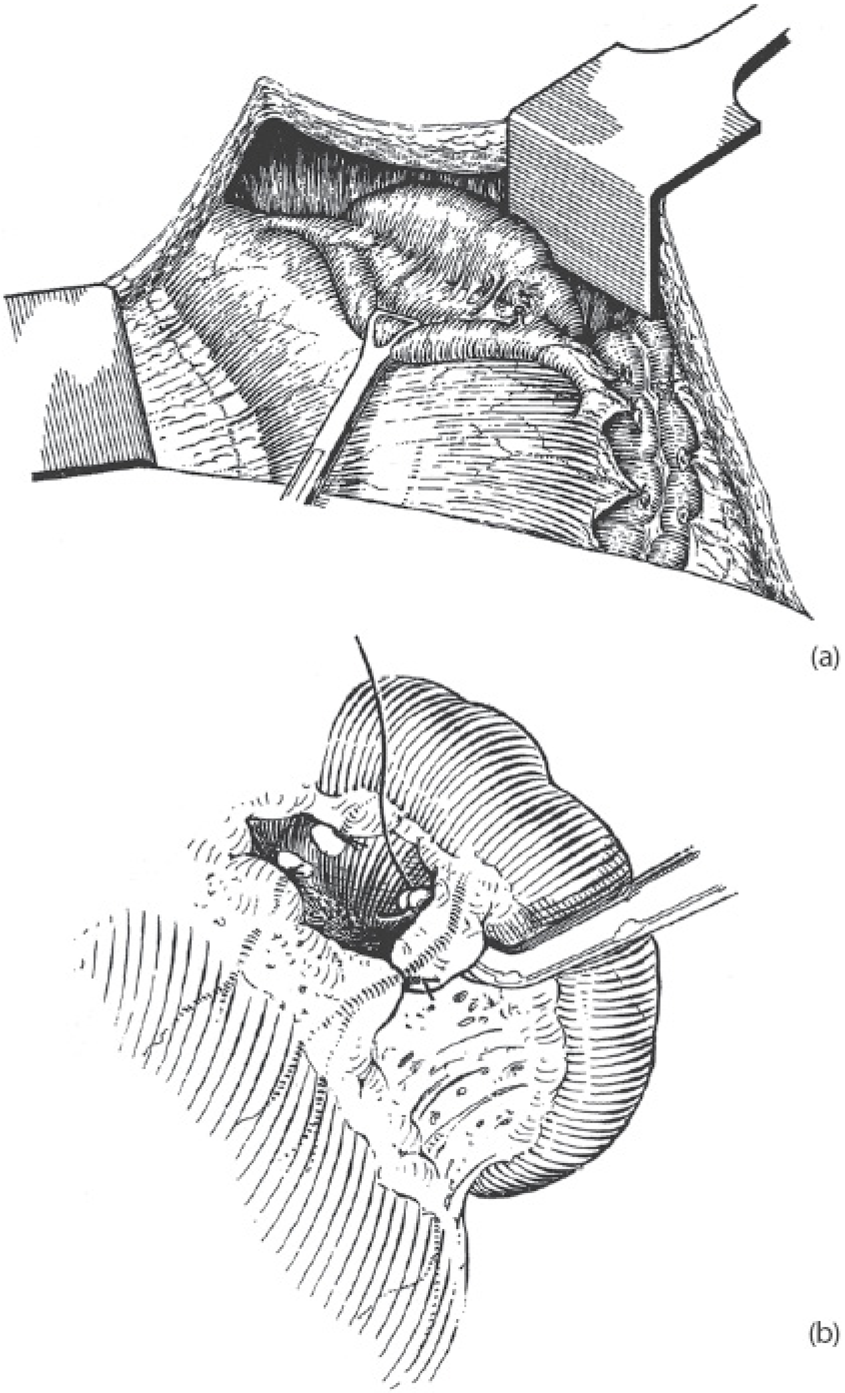
Adequate exposure of the gastroesophageal junction will usually be obtained by retracting the left lobe of the liver anterosuperiorly. Additional exposure may be attained, if necessary, by dividing the left triangular ligament in the avascular plane and then retracting the left lobe of the liver to the right (**Figure 28.14**). This maneuver is particularly useful in infants and can allow a more limited incision. The use of the self-retaining retractor is invaluable in obtaining stable exposure of the operative field.



28.14

MOBILIZATION OF THE FUNDUS OF THE STOMACH

The proximal one-third to one-half of the greater curvature of the stomach is liberated from its attachment to the spleen by ligating and dividing the upper short gastric vessels in the gastrosplenic ligament. This is facilitated by retracting the fundus rightwards with a Babcock clamp (**Figure 28.15a**). These vessels may be divided and secured in a number of ways. In infants and small children, these may simply be electrocoagulated. In older children, these vessels can be divided with an advanced energy device or ligated.

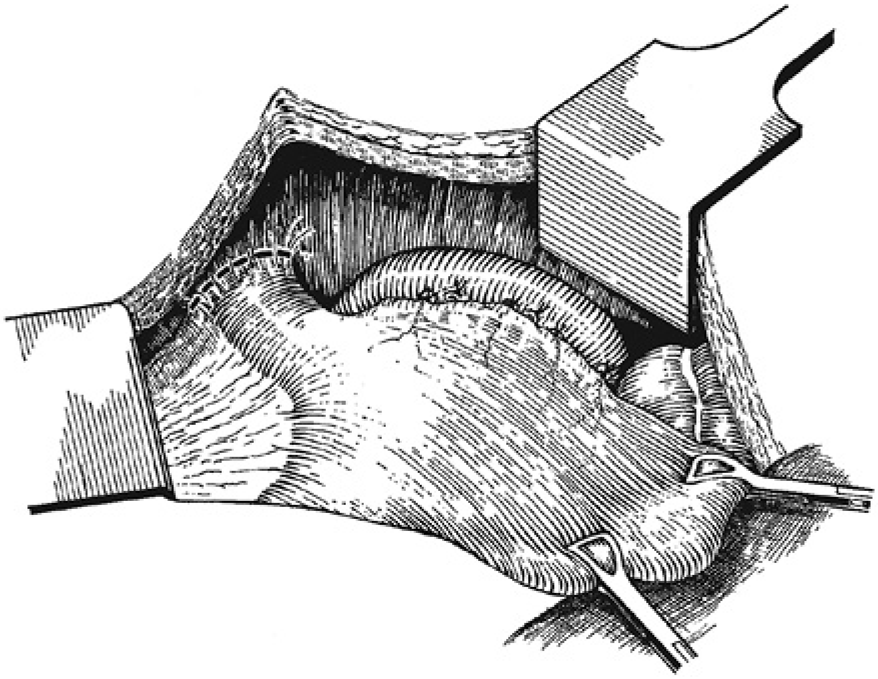


28.15a,b

When the vessels in the gastrosplenic ligament have been divided, the spleen should be allowed to fall back into the posterior peritoneum, thereby avoiding inadvertent trauma. The fundus is now sufficiently free to allow for a loose ("floppy") fundoplication (**Figure 28.15b**).

EXPOSURE OF THE ESOPHAGEAL HIATUS

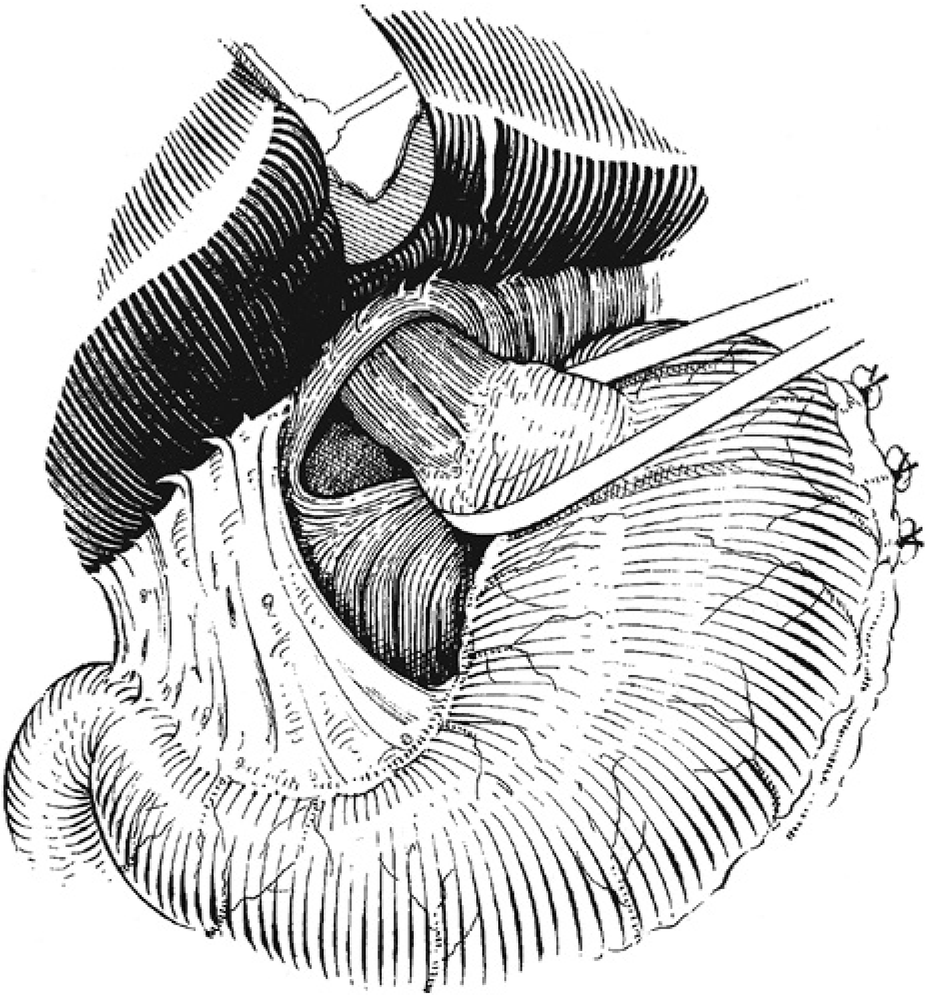
The phrenoesophageal membrane is placed on stretch by downward traction on the stomach while the diaphragmatic muscle is retracted superiorly. The avascular membrane marked by a "white line" is incised with scissors and the musculature of the esophagus displayed (**Figure 28.16**). The anterior vagus nerve will be seen coursing on the surface of the esophagus. It should be carefully protected and preserved.



28.16

MOBILIZATION OF THE DISTAL ESOPHAGUS

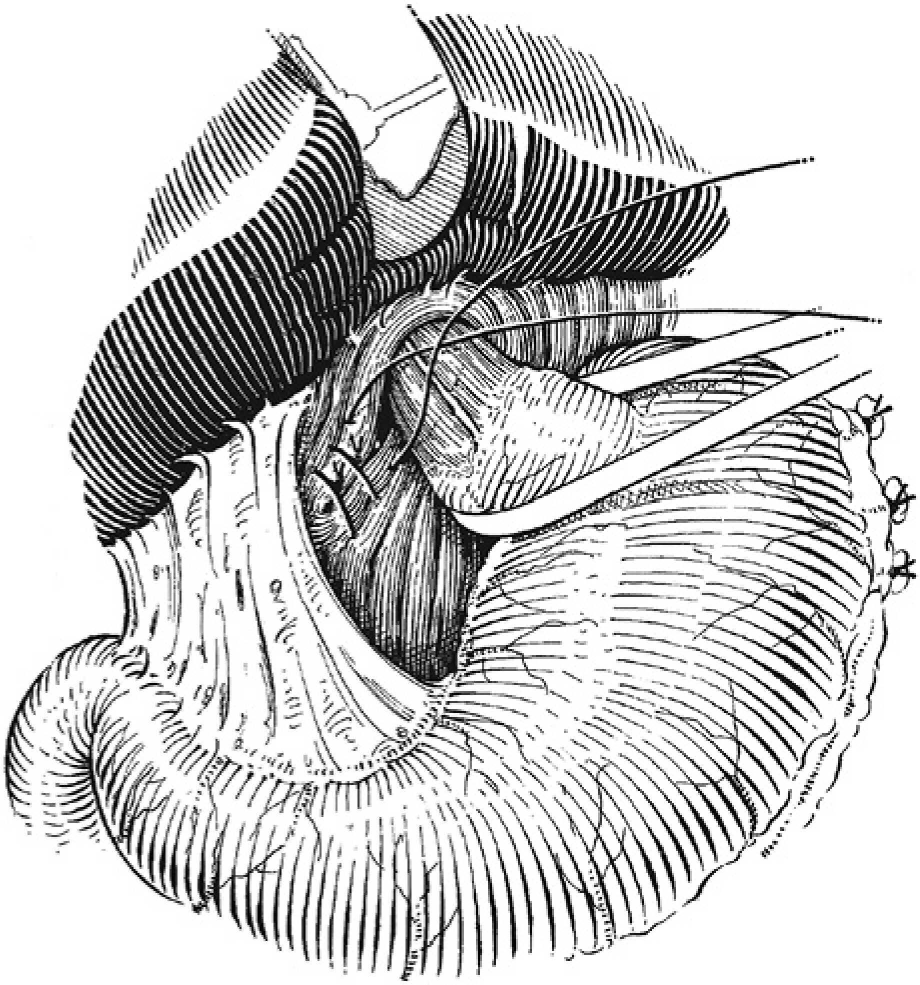
Using a combination of sharp and blunt dissection, the lower end of the esophagus is encircled, avoiding injury to the posterior vagus nerve. A rubber sling is placed around the esophagus incorporating the posterior vagus nerve, which will be included in the fundoplication (**Figure 28.17**). The lower 2–3 cm of esophagus hiatus is completely exposed by dividing the upper part of the gastrohepatic omentum above the left gastric vessels.



28.17

NARROWING OF THE HIATUS

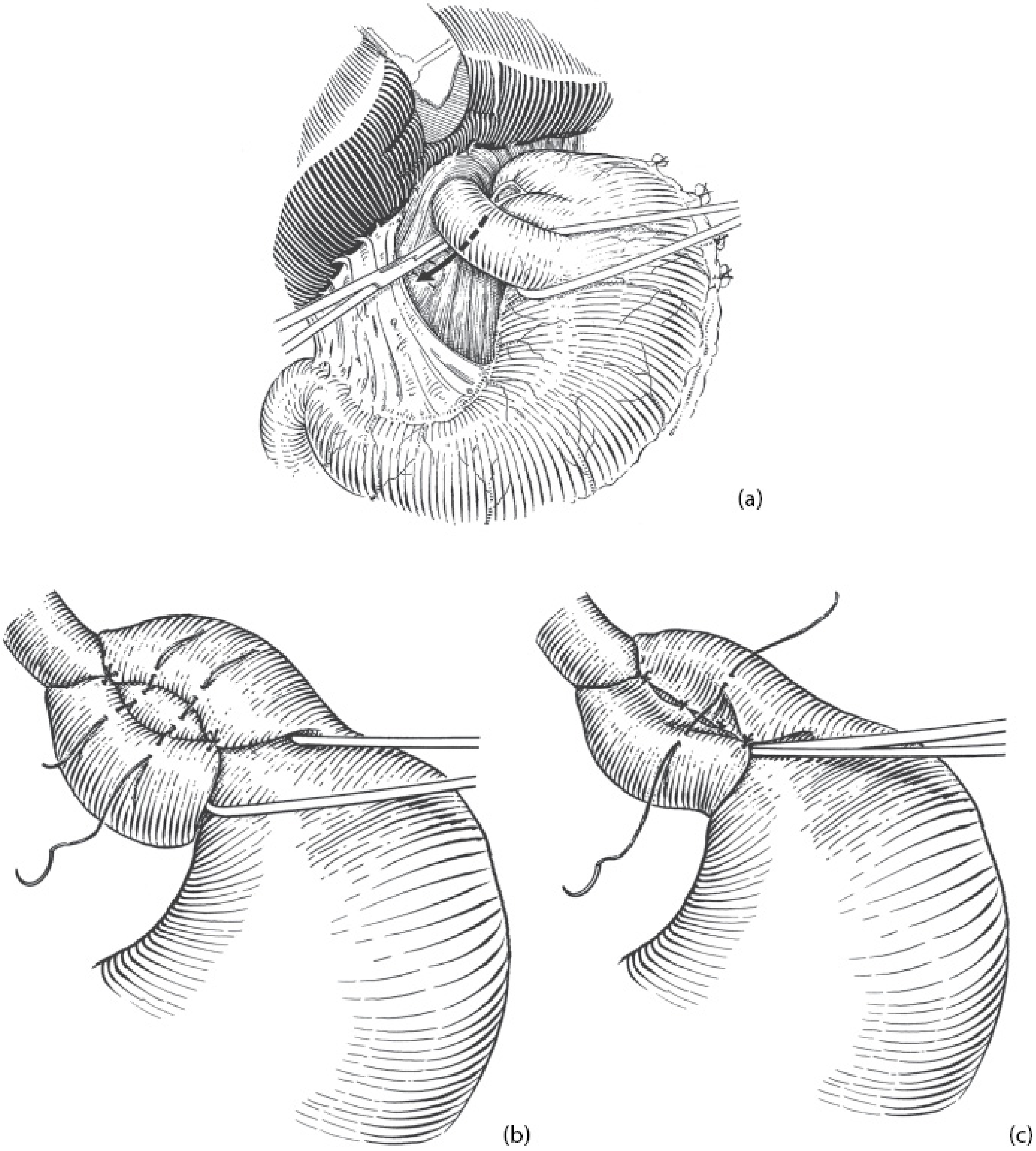
The esophageal hiatus is narrowed posterior to the esophagus by placing deep sutures through the crura of the diaphragm. The sutures are tied loosely to prevent them cutting through, but leaving sufficient space alongside the esophagus to allow the tip of a finger to pass (**Figure 28.18**). Two, three, or more sutures may be required for this purpose. In very large hiatal hernias, one or two anterior sutures may be placed to avoid kinking the esophagus by displacing it anteriorly. Some surgeons use pledgets on the crural sutures but we do not due to concern of erosion into the esophagus.



28.18

CONSTRUCTION OF THE FUNDOPLICATION

The mobilized fundus of the stomach is folded behind the esophagus so that the invaginated part of the stomach appears on the right side of the esophagus (**Figure 28.19a**). It is important not to twist the stomach during this maneuver and to ensure that the stomach has been sufficiently mobilized to be able to fashion a loose wrap.

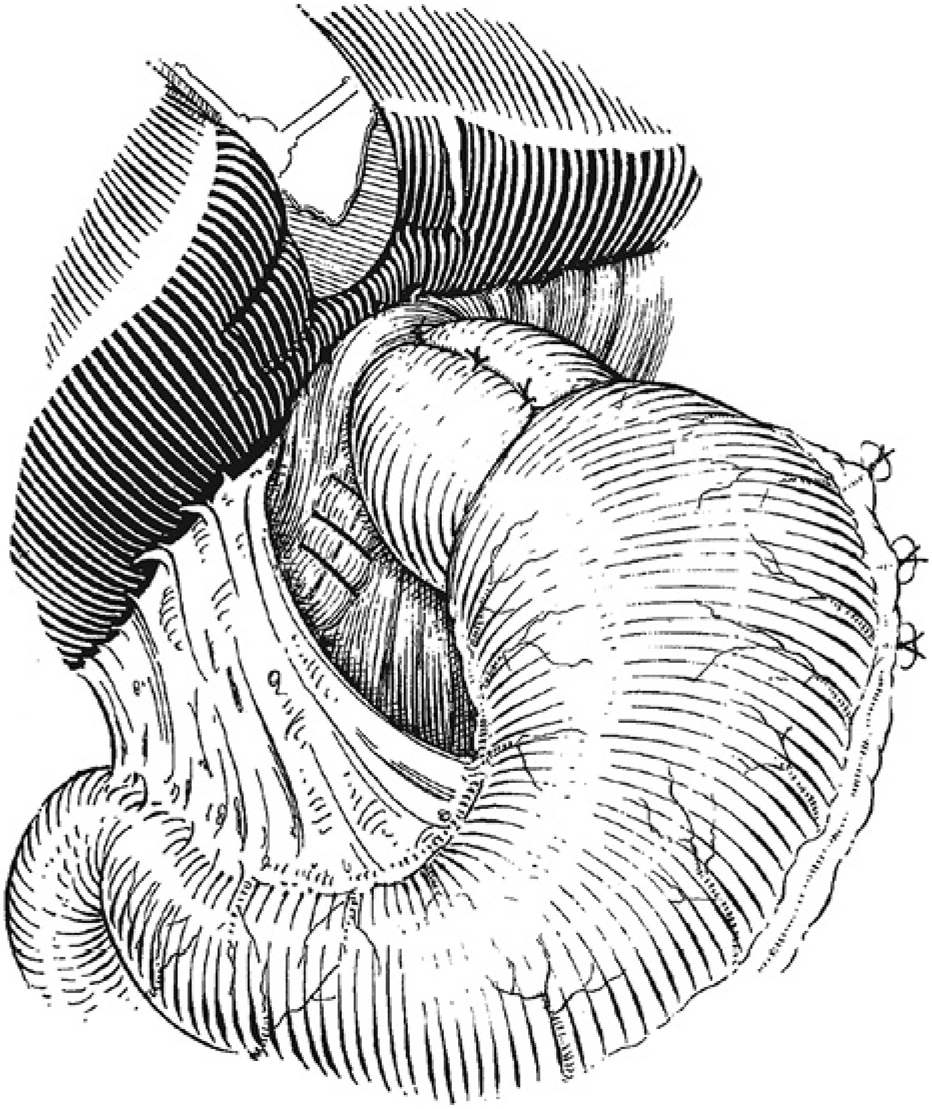


28.19a–c

The length of the wrap varies from 1.0 cm in the infant up to 2.5 cm in the older child. Commencing at the level of the gastroesophageal junction, three or four sutures of non-absorbable material (3/0 or 4/0) are placed through the stomach and esophageal muscle. Each suture passes from left to right through the anterior wall of the stomach, through the esophageal muscle (taking care not to enter the lumen of the esophagus), and through the wall of the mobilized portion of the fundus of the stomach, which has been folded behind the esophagus (**Figure 28.19b**). Traction on the first (untied) suture brings the rest of the operating field clearly into view and facilitates the insertion of the remaining two or three proximal parallel sutures through the anterior wall of the stomach, esophageal muscle, and "prolapsed" fundus. When all these sutures are in place, they are tied serially without tension on the wrap.

A second layer of non-absorbable sutures including the seromuscular surface of the stomach only may be placed superficial to the primary sutures to prevent disruption of the wrap (**Figure 28.19c**). This is done variably and is done by us only if there seems to be tension on the suture line.

The proximal suture of the second layer should be passed through the anterior wall of the esophageal hiatus in the diaphragm. Optionally, an additional two or three sutures may be placed between the hiatus and the fundoplication to prevent the wrap migrating into the posterior mediastinum (**Figure 28.20**).



28.20

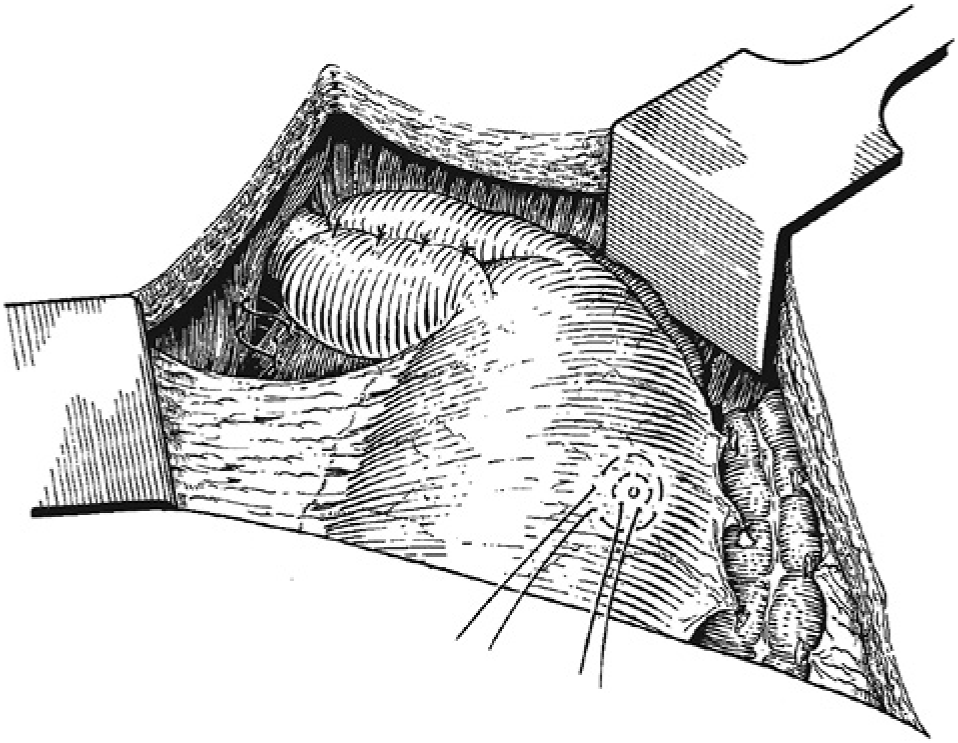
CLOSURE

The wound is closed in layers. A subcuticular suture approximates the skin edges.

GASTROSTOMY

A feeding gastrostomy should be constructed in neurologically impaired children who are unable to eat normally. A suitable area on the anterior wall of the stomach is selected to permit the gastrostomy site to be anchored to the anterior abdominal wall without exerting traction on the fundoplication. Routine gastrostomy placement for gastric decompression is not required so gastrostomies are placed only when needed for nutritional support.

Two rows of circumferentially placed non-absorbable sutures are placed through the seromuscular layer of the stomach (**Figure 28.21**). A suitable-size balloon-tip gastrostomy tube or a Malecot catheter is inserted through a centrally placed gastrotomy into the stomach and the sutures are tied, invaginating the gastrotomy site. The tube is brought out through a separate stab incision in the left hypochondrium, and the stomach wall is sutured to the anterior abdominal wall at the exit site to prevent leakage.



28.21

Postoperative care

Nasogastric decompression is used selectively based on the degree of ileus that is anticipated. In patients who do not require lysis of adhesions from prior surgery, the postoperative ileus is mild and brief. Liquid diet may be started in these patients the night of surgery. In reoperative cases, nasogastric decompression and IV fluids are continued until postoperative ileus has resolved.

Complications

- Death following this procedure is extremely uncommon in the immediate postoperative period. Almost all mortalities occur in children with complex chronic medical conditions such as neurological impairment, cardiac anomalies, and pulmonary disease. Careful monitoring and attention to pulmonary toilet is critical in these patients.
- Wound infection occasionally occurs.
- Dysphagia may result from a wrap that is either too long or too tight. If an appropriate-sized dilator is used during the operation, this should be transient, typically resolving 2–3 weeks postoperatively. Some children may require brief support with nasogastric feeding while postoperative edema resolves. Dilatation should be reserved for refractory cases as this increases the risk of recurrent gastroesophageal reflux.
- The gastroesophageal reflux may recur because of either disruption of the fundoplication or herniation of the fundoplication into the posterior mediastinum.
- Paraesophageal hernia occurs following inadequate approximation or disruption of the crural repair.
- Gas bloat, hiccup, retching, and dumping symptoms are usually transient.
- Adhesion intestinal obstruction is particularly common if an additional intra-abdominal procedure, such as gastrostomy, incidental appendectomy, or correction of malrotation is performed. It is important to alert the parents to the danger of intestinal obstruction, as the inability to vomit may result in inordinate delay in establishing the diagnosis.

VIDEO

OPS_8e_video_28.1 Laparoscopic Nissen fundoplication

FURTHER READING

Goldin AB, Sawin R, Seidel KD, Flum DR. Do antireflux operations decrease the rate of reflux-related hospitalizations in children? *Pediatrics* 2006; 118: 2326–33.

McHoney M, Wade AM, Eaton S, et al. Clinical outcome of a randomized controlled blinded trial of open versus laparoscopic Nissen fundoplication in infants and children. *Ann Surg* 2011; 254: 209–16.

Ngerncham M, Barnhart DC, Haricharan RN, et al. Risk factors for recurrent gastroesophageal reflux disease after fundoplication in pediatric patients: A case-control study. *J Pediatr Surg* 2007; 42: 1478–85.

Pacilli M, Eaton S, Maritsi D, et al. Factors predicting failure of redo Nissen fundoplication in children. *Pediatr Surg Int* 2007; 23: 499–503.

Rosen R, Vandenplas Y, Singendonk M, et al. Pediatric Gastroesophageal Reflux Clinical Practice Guidelines: Joint Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition. *J Pediatr Gastroenterol Nutr* 2018; 66: 516–54.

Srivastava R, Berry JG, Hall M, et al. Reflux related hospital admissions after fundoplication in children with neurological impairment: Retrospective cohort study. *Br Med J* 2009; 339: b4411.

St Peter SD, Barnhart DC, Ostlie DJ, et al. Minimal vs extensive esophageal mobilization during laparoscopic fundoplication: A prospective randomized trial. *J Pediatr Surg* 2011; 46: 163–8.

29

Total esophagogastric dissociation

RICCARDO COLETTA AND ANTONINO MORABITO

HISTORY

The relationship between hiatus hernia of the stomach and gastroesophageal reflux disease (GERD) was emphasized by surgeons such as Philip Allison in Leeds, UK, and Norman Barrett in London, UK, during the 1950s. Early operations were described to reduce the hernia within the abdominal cavity, restore the normal anatomical relationship of the crura and distal esophagus, and recreate the angle of His.

In 1936, the German surgeon Rudolf Nissen, while Chief of Surgery in Istanbul, improvised a 360-degree fundoplication to protect an esophagogastric anastomosis in a young man with a penetrating esophageal ulcer. He subsequently noted that the patient's symptoms of reflux were entirely eradicated. Some years later in 1955, and now back in Basel, Switzerland, he performed this electively in a 49 year-old woman. The clinical outcome was excellent and he published a description the following year.

Although widely used and clearly effective in children, the Nissen procedure does have a significant recurrence rate, particularly in those with severe neurological impairment (NI). Therefore in 1997 Adrian Bianchi from Manchester, UK, proposed total esophagogastric dissociation (TEGD) initially as a salvage procedure following failed fundoplication. More recently, it has been used as an alternative primary procedure for GERD in a selected group of NI children.

PRINCIPLES AND JUSTIFICATION

GERD may affect otherwise normal children leading to symptoms of vomiting, retrosternal pain, failure to thrive, and complications such as esophagitis, bleeding, and stricture. However, severe GERD is most usually associated with NI and is more difficult to treat than in otherwise normal children. Contributory factors include oromotor dysfunction, spasticity or hypotonicity, and intestinal dysmotility, which affect swallowing and protective reflexes and result in drooling and pooling of secretions predisposing to aspiration. Nasopharyngeal reflux is often also present due to the dysfunction of the soft palate. In these patients, recurrent respiratory problems and failure to thrive are common and respiratory deterioration is a leading cause of premature death.

Effective conservative management of GERD in NI children may not be possible and therefore they come to surgical attention for consideration of a feeding gastrostomy to avoid repeated passage of a nasogastric or nasojejunal feeding tube. Although advocated by some, placement of a gastrostomy tube alone will aggravate the GERD in well over 50% of children. If the child's overall condition does not allow for a safe concomitant antireflux procedure, a temporary transgastric jejunal feeding tube/button or a tube jejunostomy may be better prospects.

Repeated surgery is often hazardous, with increased morbidity, and these patients are at high anesthetic risk. A single operative intervention is often the preferred choice to minimize the risk of GERD and to address both feeding and respiratory problems.

TEGD with gastrostomy is designed specifically to meet the essential feeding needs of NI children by eliminating GERD and the possibility of its recurrence. It is important to remember that TEGD does not alter the pharyngeal incoordination and the potential morbidity from aspiration of saliva, nor does it resolve the troublesome centrally coordinated retching. Primary TEGD with gastrostomy should ideally be offered as a single definitive procedure since it meets the family's and caregiver's expectation. The aim is to eliminate the risk of recurrence of reflux, and hence to reduce the incidence of aspiration, chest infections, and hospitalization. Our experience has confirmed that TEGD improves nutrition and growth, and significantly improves the quality of life for both the child and the family.

Rescue TEGD is indicated as a salvage procedure following failure of conventional antireflux surgery. In this scenario, the morbidity of TEGD is increased by previous antireflux interventions, the anatomical condition of the lower esophagus, and also the higher risk of vagal injury. However, the benefits far outweigh the risks.

Experience has highlighted "additional indications" for TEGD in otherwise difficult situations, such as gastric necrosis or bleeding remnant after subtotal gastric resection where there is a need to ensure esophageal transit of food to the small bowel without clinically relevant reflux.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

Videofluoroscopy, pH studies (24 hours or impedance), radioisotope scans, and contrast studies are used to confirm the diagnosis of severe GERD. Delayed gastric emptying and the possible presence of an esophageal stenosis or ulceration are relevant to the surgeon, and a review of lung function is significant to the anesthetist.

The involvement of a pediatric respiratory physician and a pediatric intensivist is appropriate to ensure that the child is in their best possible condition for surgery. Parenteral nutrition could be employed if nutritional status is a concern.

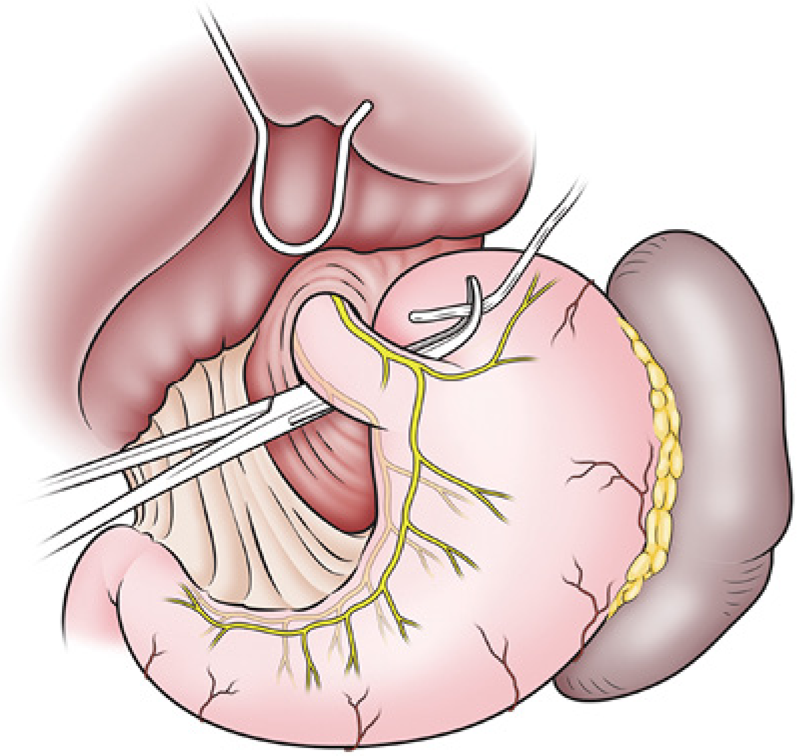
A multidisciplinary team should address all the needs of the individual patient and their family prior to surgery. It is the surgeon's obligation to become personally involved with the family or caregivers through in-depth counselling sessions such that the family's concerns and expectations are fully addressed. The patient's family must be fully aware that TEGD is designed to eliminate reflux and food aspiration, improve lung function, and secure better nutrition and growth. This procedure will not address the swallowing difficulty, saliva aspiration, and the centrally mediated "retching" that are often so distressing to the family. For this reason an otolaryngologist is often consulted to assess the patient prior to surgery.

OPERATION

The TEGD technique described is that proposed originally by Bianchi in 1997. Recently, laparoscopic and robot-assisted TEGD have been reported but more experience is needed using these approaches.

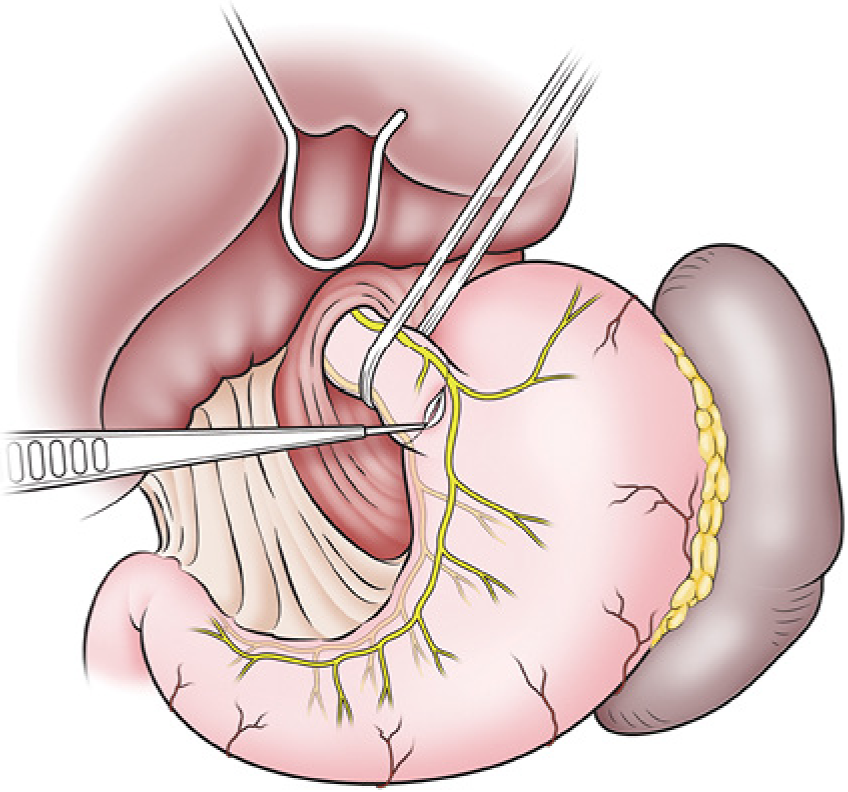
The technique uses an upper abdominal approach made via a midline incision.

Adequate exposure of the gastroesophageal structure is achieved by gentle retraction of the left lobe of the liver anterosuperiorly (**Figure 29.1**). This maneuver allows liberal mobilization of the esophagus at the hiatus of the diaphragm. In older children, particularly those with kyphoscoliosis or who are obese, the use of a self-retaining retractor is a possible option to improving exposure.



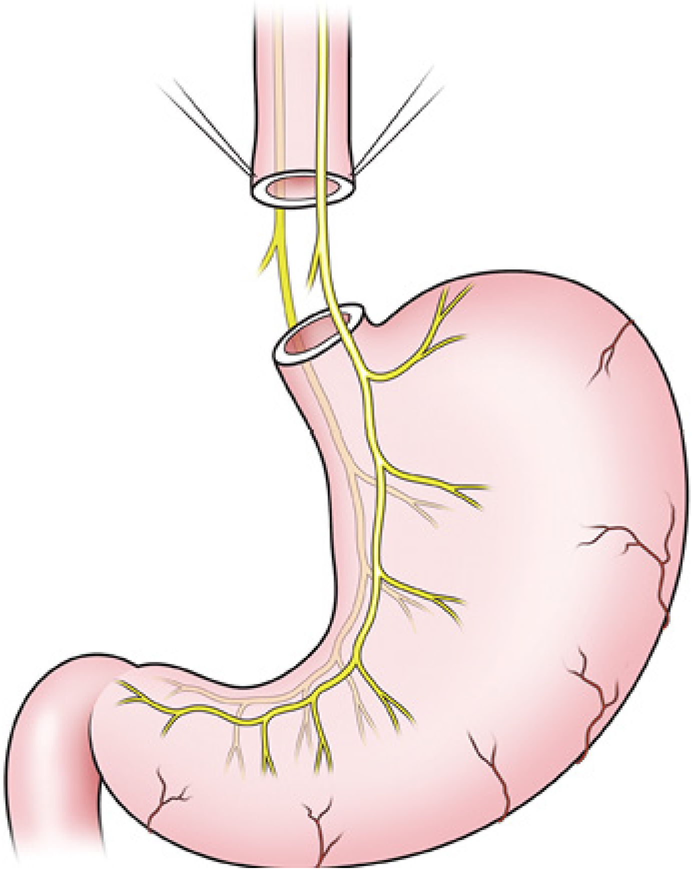
29.1

The posterior vagus nerve is protected, the esophagus is detached at the cardioesophageal junction, and the stomach is oversewn (**Figure 29.2**). In case of rescue TEGD, any residual fundal wrap from a previous Nissen procedure is unraveled, and the subdiaphragmatic esophagus is mobilized preserving any pre-existing gastrostomy.



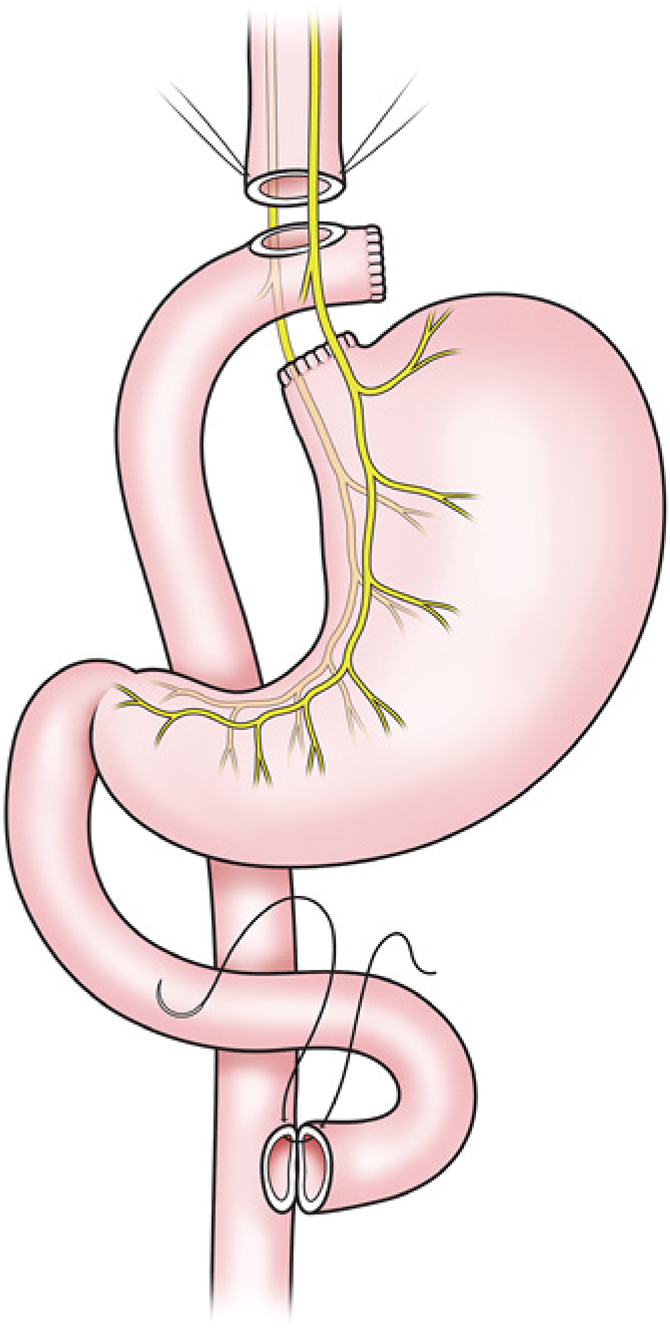
29.2

Two stay sutures in 3/0 silk are located at the extremities of the distal esophagus (**Figure 29.3**). The distal esophageal stump is resected at the stomach level with a bipolar to avoid reflux. The stomach is closed in double layer using a running suture with 5/0 Maxon™.



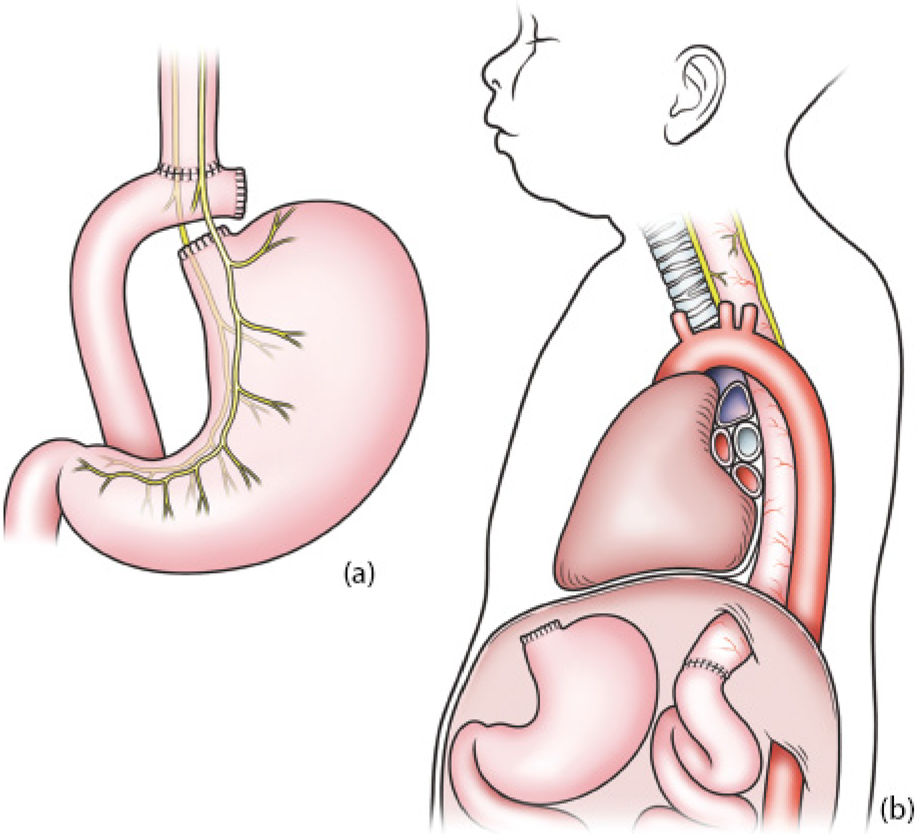
29.3

The jejunum is divided and the distal end is passed in Roux-en-Y isoperistaltic fashion on a tension-free mesentery through the transverse mesocolon and behind the stomach (**Figure 29.4**). The distal end of the jejunum is anastomosed with the esophagus at the level of the diaphragm.



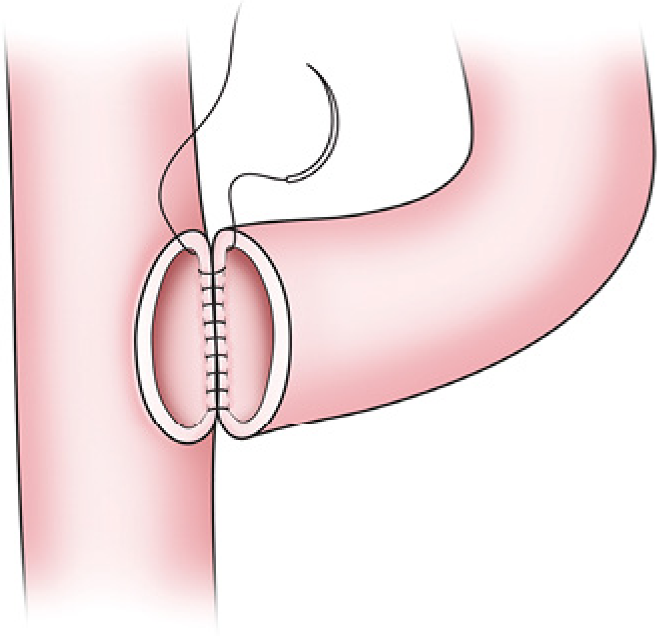
29.4

A “wide oblique” end-to-side rather than a circular end-to-end anastomosis is performed between the distal esophagus and the jejunum using absorbable sutures (4/0 or 5/0 Maxon) (**Figure 29.5a**). The oblique end-to-side anastomosis reduces/minimizes any possibility of stenosis and is also performed to avoid stretching of the mesentery that could compromise blood supply. The mesenteric window in the mesocolon and the small bowel mesentery are approximated in 5/0 Maxon and the jejunum is included in the suture line (**Figure 29.5b**).



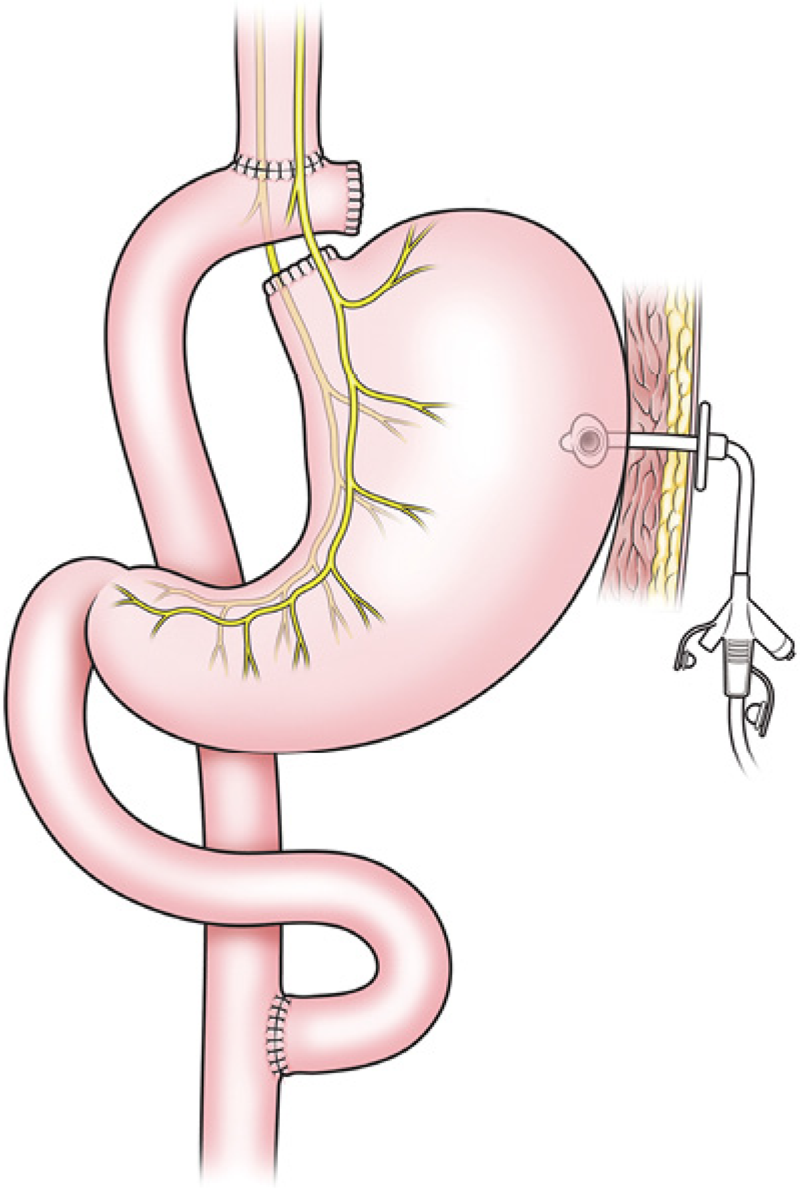
29.5a,b

Bowel continuity is established by end-to-side jejunojejunostomy approximately 40 cm distal to the esophageal anastomosis using inverted vertical mattress suture with an absorbable monofilament (5/0 Maxon) (**Figure 29.6**) and also a transanastomotic tube is inserted. However, a 20–30 cm Roux-en-Y from the esophagojejunal anastomosis is usually adequate in the pediatric population.



29.6

The operation ends with the creation of a Stamm gastrostomy (**Figure 29.7**). Double purse-string sutures are placed on the body of the stomach. The opening will be at the level of the splenic hilum. A de Pezzer or Malecot catheter is introduced while the sutures are elevated. The stomach is secured to the abdominal wall and the catheter end is grasped from a counter-incision and the tube is brought out. A Heineke-Mikulicz pyloroplasty with 4/0 or 5/0 Maxon is only performed if there has been an accidental intraoperative injury to the vagus nerve or in the event of rescue TEGD. The abdominal wall is closed in layers with polyfilament absorbable suture (3/0 and 4/0 Vicryl^{TM}). The skin is approximated in Biosyn^{TM} 5/0 and skin glue.



29.7

POSTOPERATIVE CARE

In view of their precarious respiratory and physical state, these children benefit from a brief postoperative period of assisted ventilation and intensive care management. The stomach is drained continuously postoperatively through the gastrostomy and the transanastomotic tube is aspirated every 4 hours until return of bowel activity. Recovery is usually uneventful and gastrostomy feeds and oral intake commence once bowel function becomes established, commonly within 3–5 days via the transanastomotic tube.

COMPLICATIONS

In a recent review study on outcomes of TEGD in children (n = 180), early complications were observed in 16%. **Table 29.1** depicts the most common complications. The overall mortality reported in the literature is 20%, reflecting the sense of palliation in many of these children.

Table 29.1 **Possible complications: The Manchester experience**

Early	Late
Enterocutaneous fistula	Strangulation of transhiatal herniation of stomach fundus requiring partial gastrectomy
Cecal volvulus	Small bowel herniation under Roux loop with extensive necrosis
Iatrogenic diaphragmatic hernia	Paraesophageal hernia
Esophagojejunal leak	Gastrostomy dislodgement with incorrect replacement
Slow gastric emptying	Small bowel obstruction
Subphrenic collection	Esophagojejunal stricture
Small bowel obstruction	Paraesophageal hernia
Intra-abdominal collection	Bile reflux
Necrosis of Roux-en-Y loop	Small bowel herniation into chest
	Paraoesophageal hernia

The data are similar to the authors' experience of primary TEGD (n = 49) and rescue TEGD (n = 17) performed, which were pyloroplasty leak (n = 1), esophagojejunal anastomosis leak (n = 3), wound dehiscence (n = 2), and left subphrenic collection (n = 1) (overall 14% complication rate). Late complications experienced in the authors' series were esophagojejunal anastomotic stenosis (n = 1) successfully managed with dilatation, and second-look surgery in three children due to gastric volvulus (n = 1) and small bowel herniation beneath the jejunal Roux loop (n = 2).

In the authors' series the mortality rate was 7% (one child died from intra-abdominal sepsis post-esophagojejunal anastomotic breakdown and four children died due to underlying conditions such as further deterioration of their neurological condition, chronic lung disease, congenital heart disorder, and hypovolemic shock secondary to severe diarrhea and vomiting).

OUTCOME

In-hospital mortality from the TEGD procedure has been reported in literature as 0.6% with no deaths within the first 30 days. This is similar to the overall in-hospital mortality rate (1.3%) of 9987 children who underwent an antireflux procedure in the United States between 1996 and 2003.

The mean time to death following TEGD is 21 months (range 1–79 months). Long-term mortality rates following fundoplication range from 19% to 33% with the highest rates found among NI children. The 5-year survival rate after fundoplication for children with cerebral palsy and gastrostomy is around 59%.

Median follow-up is 13 months (range 1–100 months) over the past 10 years of TEGD in our institution. All children have had resolution of their reflux, 13% experienced bloating or pain on feeding, and 26% experienced retching unrelated to GERD. One child has self-induced vomiting, one child has colonic dysmotility, and one child has Barrett's esophagitis. There were eight late deaths related to pre-existing comorbidities.

In conclusion, TEGD is a safe definitive solution for GERD because it eliminates all risk of recurrent reflux. The authors therefore feel that TEGD can be used as a primary treatment of choice for severely neurologically impaired patients who are experiencing GERD and are completely dependent on tube feeds.

FURTHER READING

Bianchi A. Total esophagogastric dissociation: An alternative approach. *J Pediatr Surg* 1997; 32: 1291–4.

Buratti S, Kamenwa R, Dohil R, et al. Esophagogastric disconnection following failed fundoplication for the treatment of gastroesophageal reflux disease (GERD) in children with severe neurological impairment. *Pediatr Surg Int* 2004; 20: 786–90.

Dall'Oglio L, Gatti C, Villa M, et al. A new and successful chance in surgical treatment of gastroesophageal reflux in severely neurologically impaired children: Bianchi's procedure. *Eur J Pediatr Surg* 2000; 10: 291–4.

Danielson PD, Emmens RW. Esophagogastric disconnection for gastroesophageal reflux in children with severe neurological impairment. *J Pediatr Surg* 1999; 34: 84–6.

de Lagausie P, Bonnard A, Schultz A, et al. Reflux in esophageal atresia, tracheoesophageal cleft, and esophagocoloplasty: Bianchi's procedure as an alternative approach. *J Pediatr Surg* 2005; 40: 666–9.

Fonkalsrud EW, Bustorff-Silva J. Interposition of a jejunal segment between esophagus and pylorus for treatment of multirecurrent gastroesophageal reflux. *J Pediatr Surg* 1999; 34: 1563–6.

Gatti C, di Abriola GF, Villa M, et al. Esophagogastric dissociation versus fundoplication: Which is best for severely neurologically impaired children? *J Pediatr Surg* 2001; 36: 677–80.

Goyal A, Khalil B, Choo K, et al. Esophagogastric dissociation in the neurologically impaired: An alternative to fundoplication? *J Pediatr Surg* 2005; 40: 915–19.

Lall A, Morabito A, Dall'Oglio L, et al. Total oesophagogastric dissociation: Experience in 2 centres. *J Pediatr Surg* 2006; 41: 342–6.

Lansdale N, McNiff M, Morecroft J, et al. Long-term and "patient-reported" outcomes of total esophagogastric dissociation versus laparoscopic fundoplication for gastroesophageal reflux disease in the severely neurodisabled child. *J Pediatr Surg* 2015; 50: 1828–32.

Morabito A, Lall A, Lo Piccolo R, et al. Total esophagogastric dissociation: 10 years' review. *J Pediatr Surg* 2006; 41: 919–22.

30

Achalasia

TIMOTHY D. KANE AND BENNO URE

HISTORY

Willis first described achalasia in 1672. He treated the patient by fashioning a rod out of a whalebone with a sponge on the end with which the patient was able to force food into his stomach. In 1877, Zenker and von Ziemssen, and in 1884 Mackenzie, suggested that achalasia was due to diminished contractile power of the esophageal musculature. In 1888, Meltzer and Mikulicz independently postulated that spasmodic contraction of the cardiac sphincter was the etiologic factor. In the same year, Einhorn proposed that the condition was due to failure of relaxation of the cardia upon swallowing.

PRINCIPLES AND JUSTIFICATION

Achalasia is a motility disorder of the esophagus characterized by an absence of peristalsis and a failure of relaxation of the lower esophageal sphincter. The cardinal symptoms in childhood are vomiting, dysphagia, chest pains and recurrent respiratory infections, and weight loss. The child learns to eat very slowly and to drink large quantities of fluid to encourage food to enter the stomach. At first, there is only regurgitation of food, but later vomiting of undigested food eaten days earlier occurs. The child with achalasia is often first referred to a psychiatrist for treatment of food aversion or anorexia.

Histopathology

Strips of muscle from the distal esophagus reveal varying pathologies from complete absence of ganglion cells to chronic inflammatory changes through to normal ganglia. Histochemistry reveals a significant reduction in all neuropeptides, particularly vasoactive intestinal polypeptide, galanin, and neuropeptide Y.

Treatment

MEDICAL TREATMENT

Transient relief of symptoms can be achieved with nifedipine, a calcium antagonist that reduces the pressure at the lower esophageal sphincter.

FORCEFUL DILATATION

The aim of this treatment is to physically disrupt the muscle fibers of the lower esophageal sphincter by means of pneumatic or balloon dilatation. A fluid-filled (Plummer) or air-filled (Browne–McHardy, Rider–Moller, angioplasty catheter) bag of fixed diameter, or balloon dilator (controlled radial esophageal, CRE), is positioned under fluoroscopic guidance in the distal esophagus and gently inflated. Relief of symptoms in children is at best temporary, but may occasionally last for prolonged periods. It has been shown that botulinum toxin injected into the lower esophageal sphincter musculature results in symptomatic relief, but the effect is short-lived.

SURGICAL TREATMENT

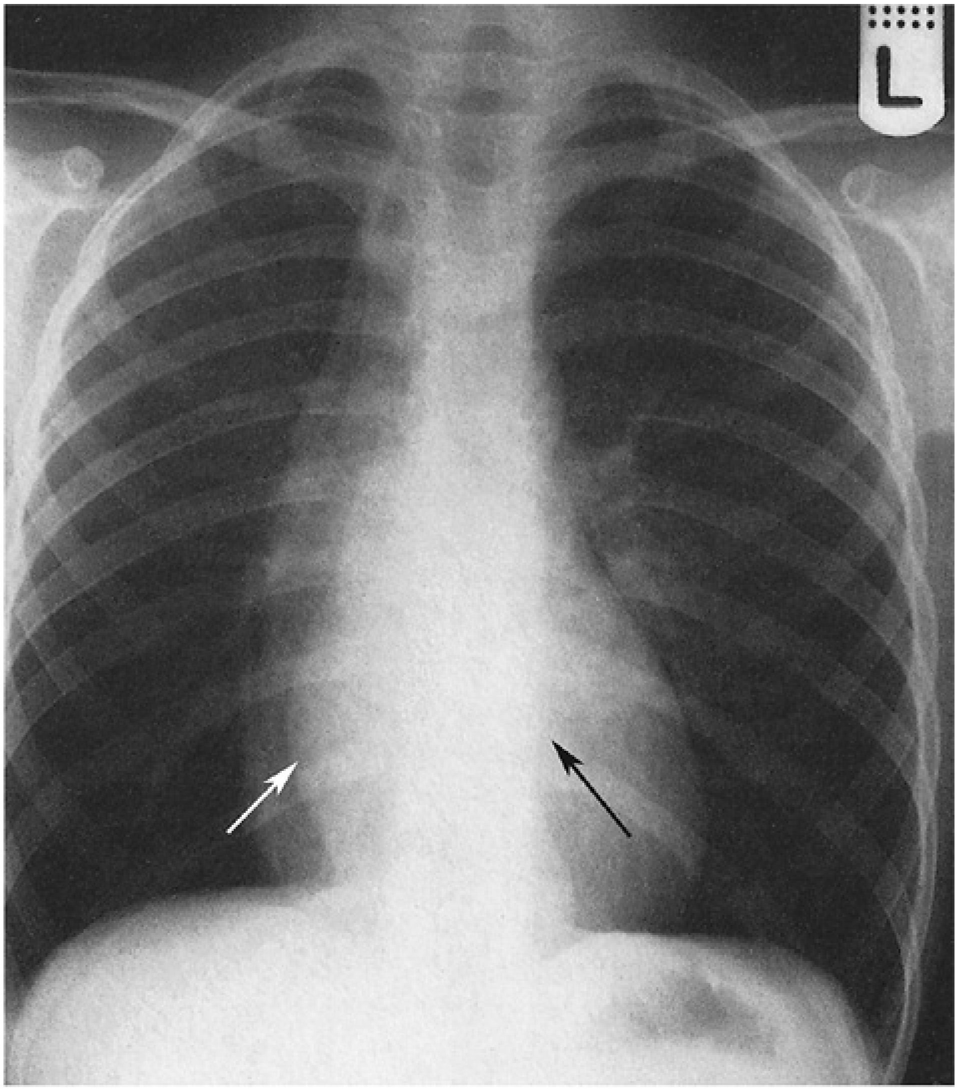
The basis of all surgical procedures for achalasia is the cardiomyotomy described in 1914 by Heller. Common controversies involve the length of the myotomy, the extent to which the myotomy extends onto the stomach, and the necessity for an antireflux procedure. It would be rare today for an open Heller myotomy to be the preferred approach for Heller myotomy for achalasia, but the historical perspective and description of the open operation may offer insight into the pathology and operative approach which may be useful in understanding the condition of achalasia.

The principle of the procedure is to perform a myotomy over the distal 4–6 cm of esophagus, extending the incision for at least 1 cm onto the anterior wall of the stomach. The myotomy is covered by a short, floppy Nissen or Dor fundoplication, or posterior Toupet fundoplication to protect against subsequent gastroesophageal reflux.

DIAGNOSIS

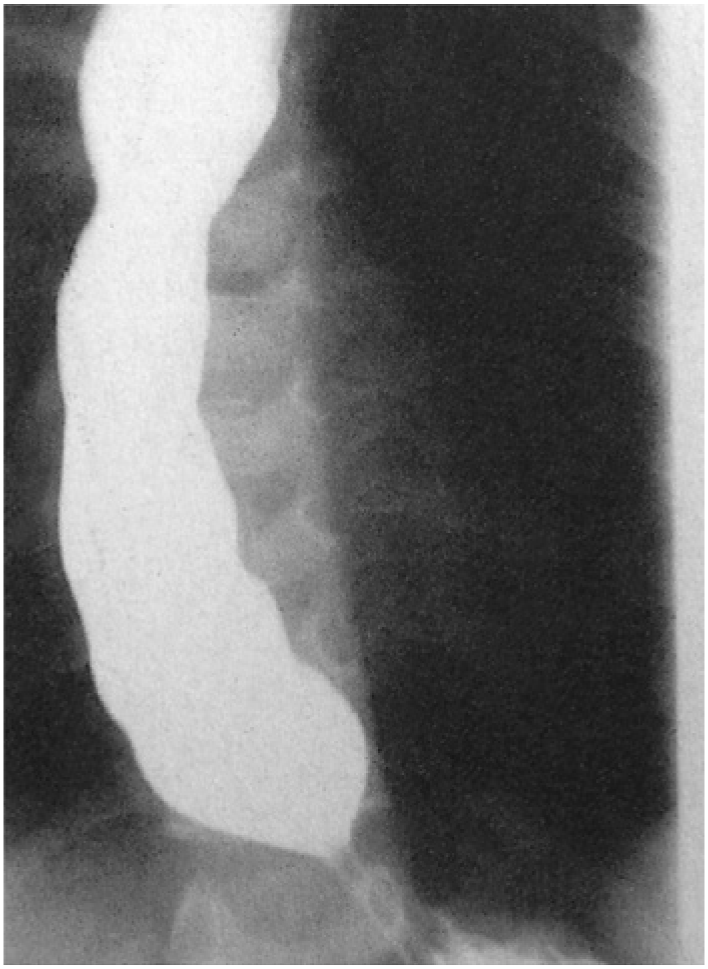
Radiologic features

A plain chest X-ray may show a dilated, food-filled esophagus with an air–fluid level. There may be radiologic signs of recurrent aspiration pneumonitis (**Figure 30.1**).



30.1

The diagnostic features of achalasia on barium swallow are a dilated esophagus, absence of stripping waves, uncoordinated contraction, and obstruction at the gastroesophageal junction with prolonged retention of barium in the esophagus. Failure of relaxation of the lower esophageal sphincter gives rise to the classical "birdbeak" deformity of funneling and narrowing of the distal esophagus (**Figure 30.2**).



30.2

Endoscopy

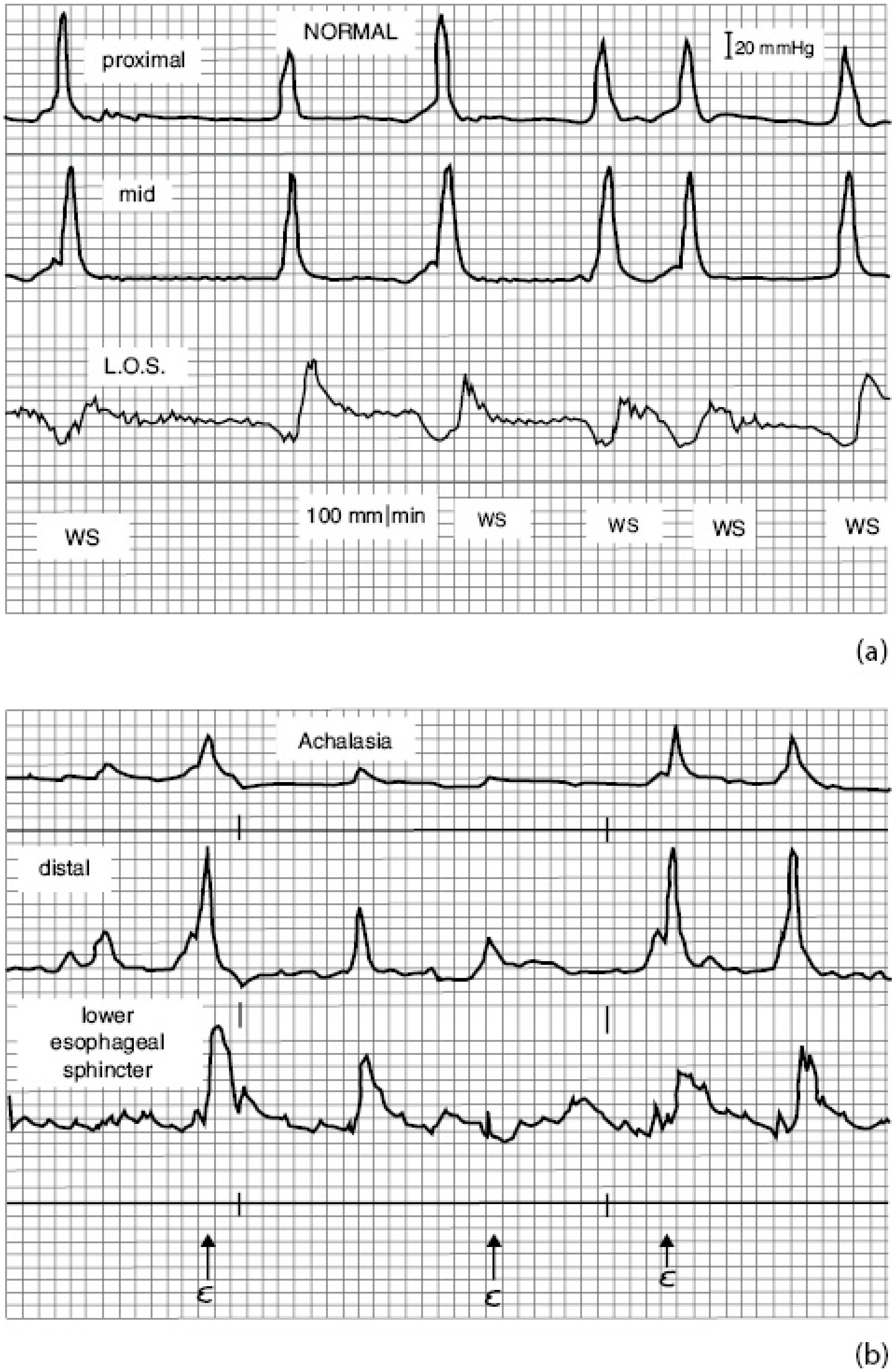
The main value of esophagoscopy is to exclude an organic cause for the obstruction.

Esophageal manometry

The criteria for diagnosis include the following (**Figure 30.3a and b**):

- High-pressure (>30 mmHg) lower esophageal sphincter zone
- Failure of the lower esophagus to relax in response to swallowing

- Absence of propulsive peristalsis
- Uncoordinated tertiary contractions in the body of the esophagus



30.3a,b

OPEN MYOTOMY

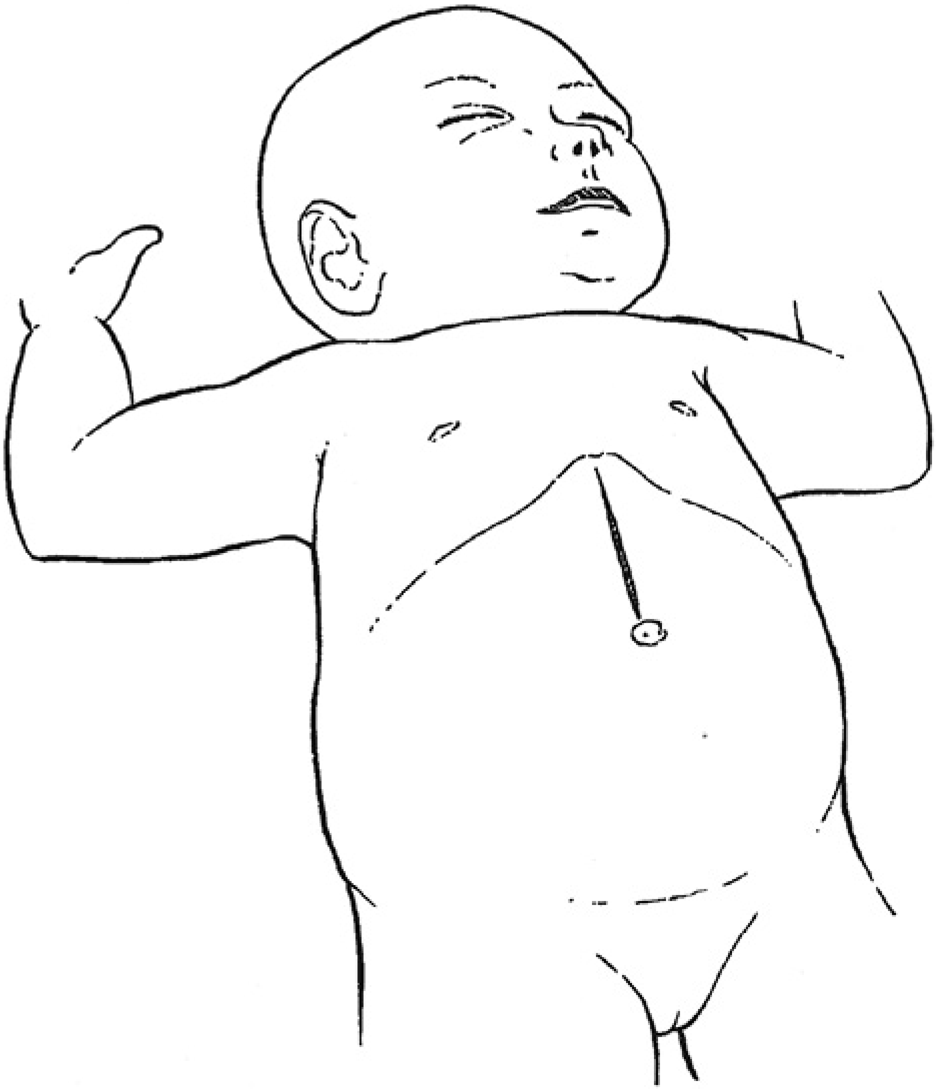
Anesthesia

General endotracheal anesthesia is administered, with the patient supine on the operating table. Measures must be taken to avoid aspiration of esophageal contents during the induction of anesthesia. Preoperative esophagoscopy is recommended to ensure complete evacuation of retained food and secretions from the esophagus. A medium-caliber nasogastric tube is passed into the stomach.

Operation

INCISION

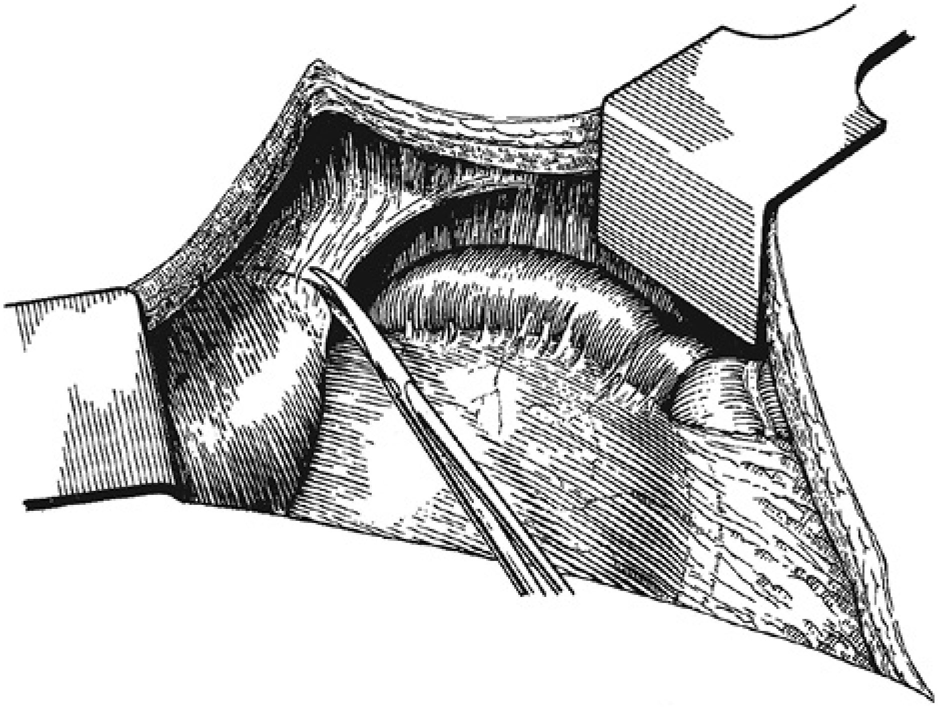
The approach is via an upper abdominal midline incision extending from the xiphisternum to the umbilicus (**Figure 30.4**).



30.4

EXPOSURE

In most cases, adequate exposure of the abdominal esophagus can be obtained by retracting the left lobe of the liver anterosuperiorly with a wide retractor (**Figure 30.5**). If necessary, additional exposure may be attained by dividing the left triangular ligament in the avascular plane and retracting the left lobe of the liver toward the midline.

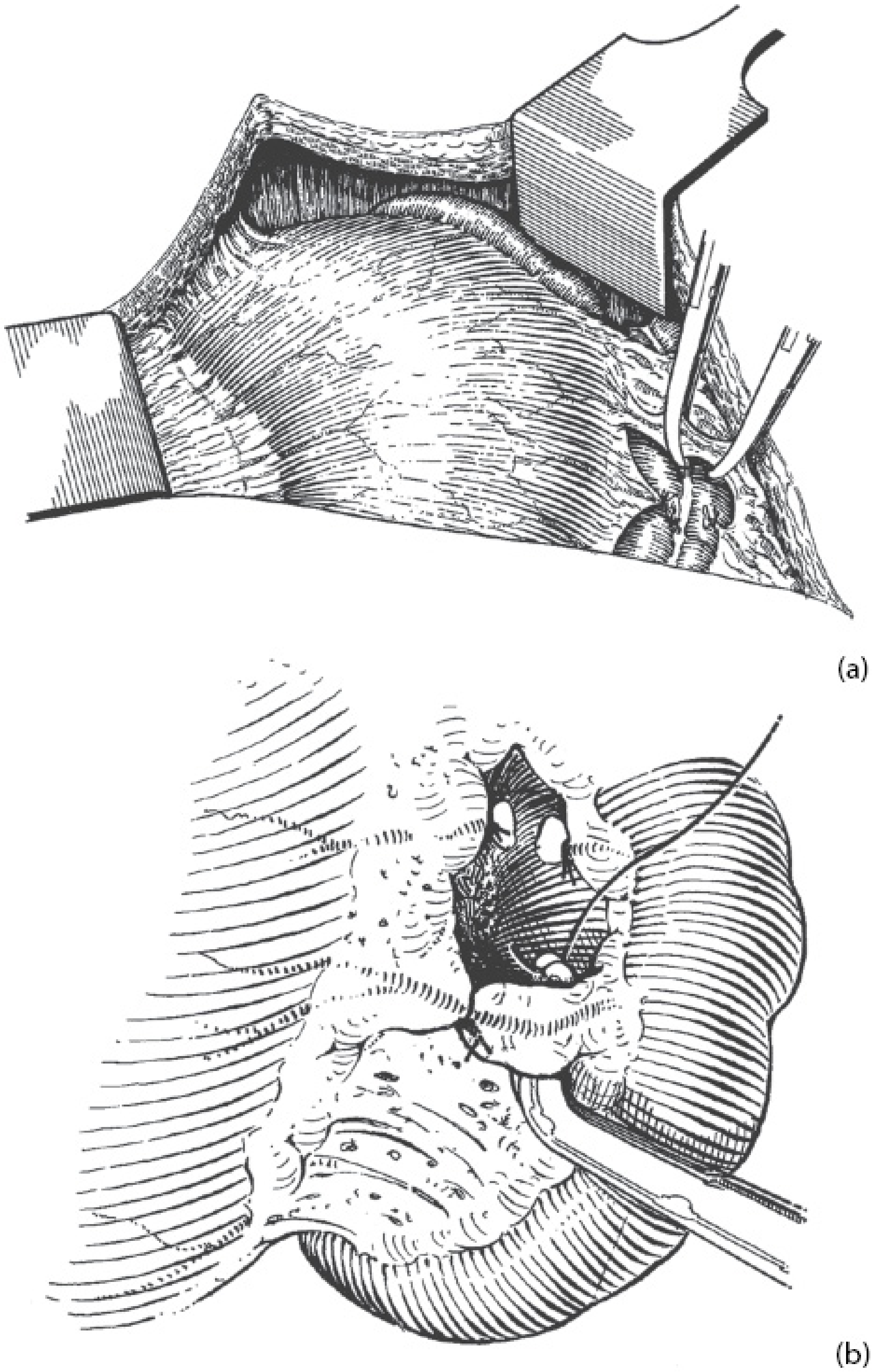


30.5

MOBILIZATION OF THE FUNDUS OF THE STOMACH

As a Nissen, Dor, or Toupet fundoplication will be performed in addition to the extended gastroesophageal myotomy, the operative procedure for fundoplication should be followed in the early stages of the operation.

The proximal one-third of the greater curvature of the stomach is liberated from its attachment to the spleen by ligating or coagulating with bipolar diathermy and dividing the short gastric vessels in the gastrosplenic ligament. This is accomplished most safely using a right-angled forceps passed around each vessel in turn or by using a sealing energy device (**Figure 30.6a**). When the vessels in the upper part of the gastrosplenic ligament have been divided, the spleen should be allowed to fall back into the posterior peritoneum, thereby avoiding inadvertent trauma. Splenectomy should never be necessary in this procedure. The fundus is now sufficiently free to allow for a loose (floppy) fundoplication (**Figure 30.6b**). The esophageal hiatus is completely exposed by dividing the upper part of the gastrohepatic omentum above the left gastric vessels.



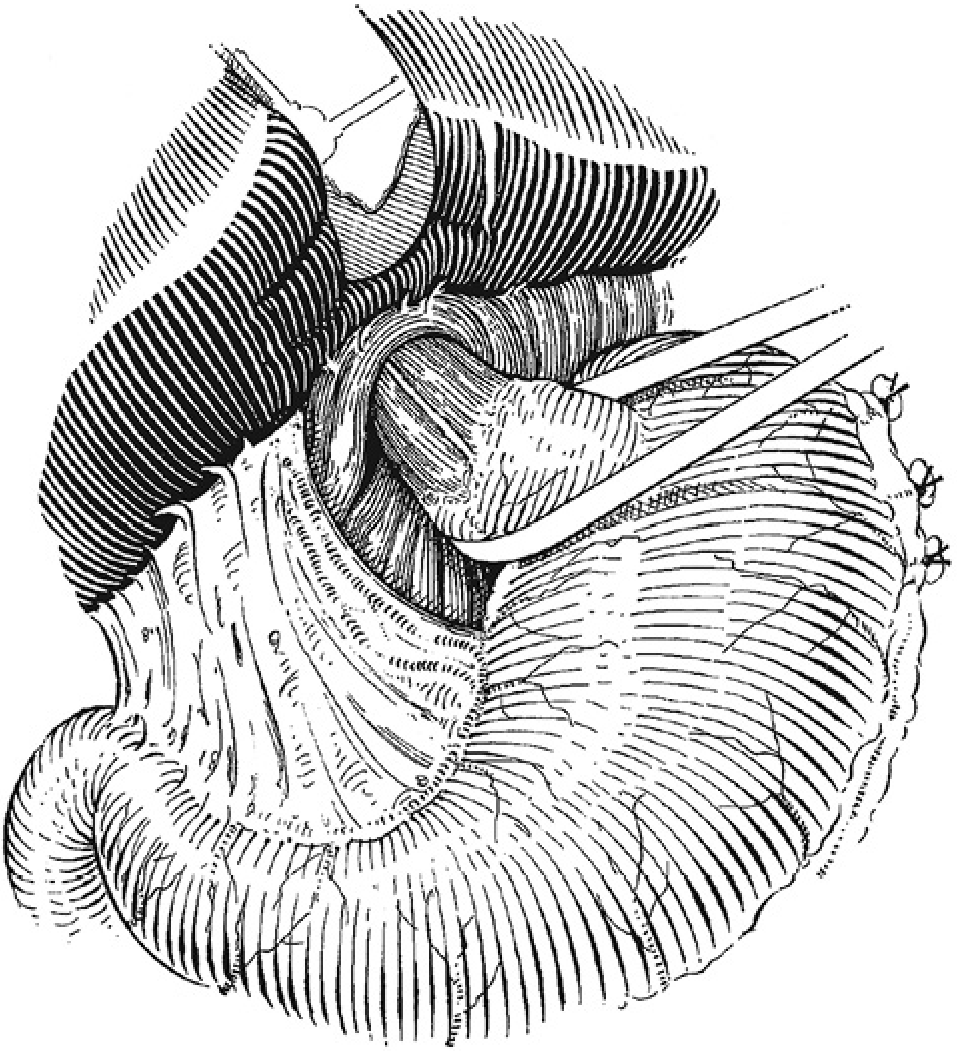
30.6a,b

EXPOSURE OF THE ESOPHAGEAL HIATUS

The phrenoesophageal membrane is placed on stretch by downward traction on the stomach while the diaphragmatic muscles are retracted superiorly. The avascular membrane is incised with scissors and the musculature of the esophagus displayed. The anterior vagal nerve will be seen coursing on the surface of the esophagus; it should be carefully protected and preserved.

MOBILIZATION OF THE DISTAL ESOPHAGUS

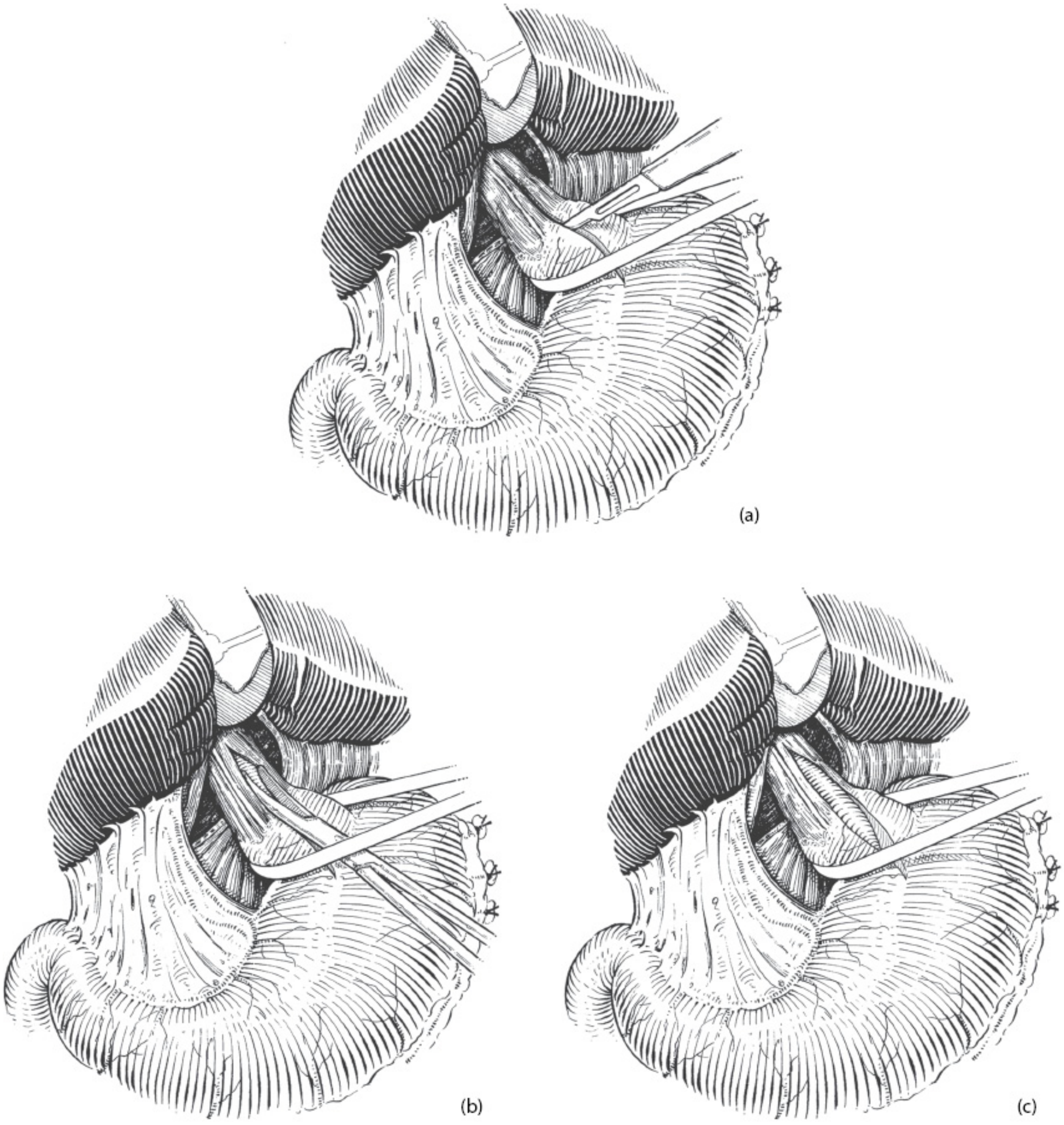
Using a combination of sharp and blunt dissection, the lower end of the esophagus is encircled, taking care not to injure the posterior vagal nerve. A rubber sling is placed around the esophagus (**Figure 30.7**). The lower 5–8 cm of esophagus is now exposed through the esophageal hiatus into the posterior mediastinum using blunt dissection with either a moist pledget or right-angled forceps.



30.7

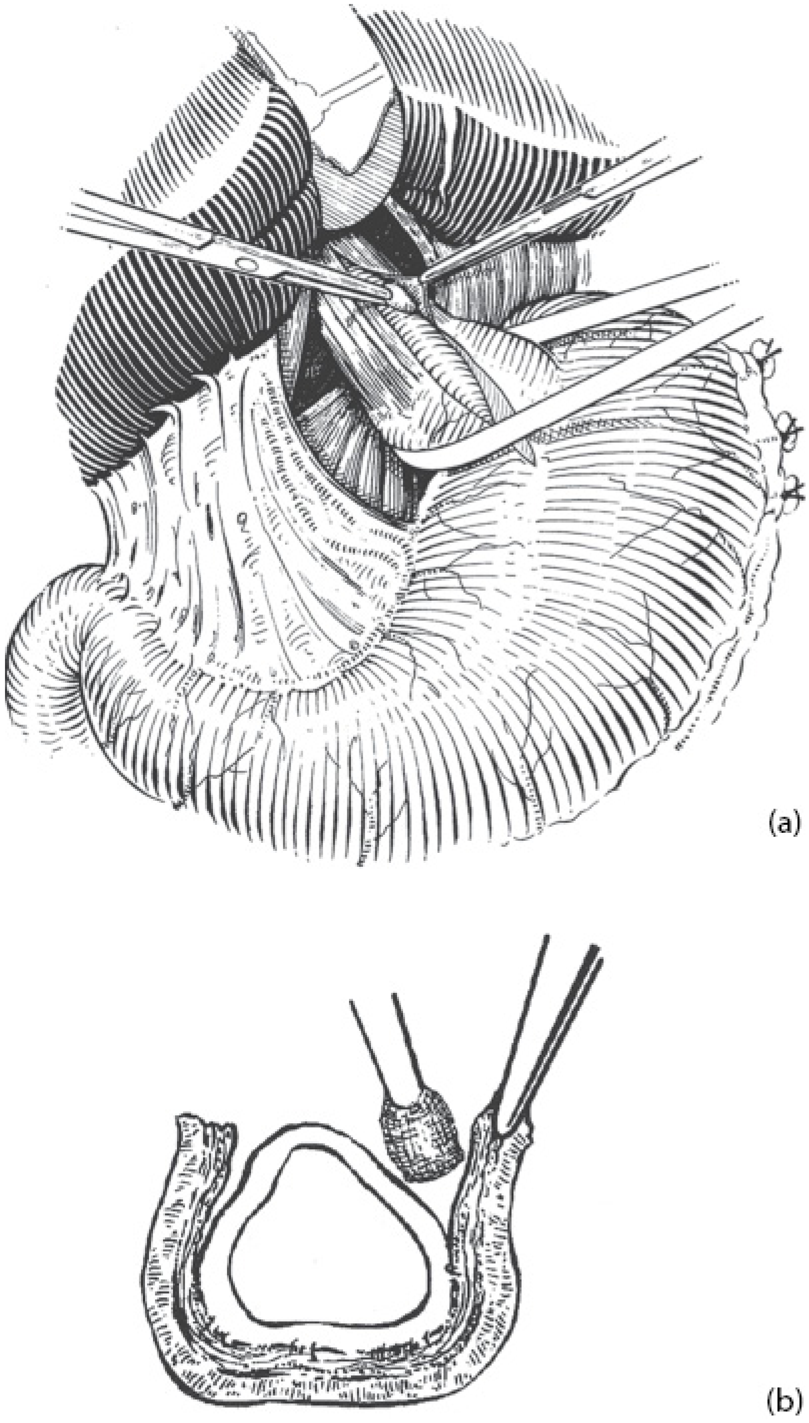
GASTROESOPHAGEAL MYOTOMY

The myotomy is performed on the anterior wall of the esophagus, extending for 1 cm onto the fundus of the stomach (as shown in **Figure 30.8a–c** to the patient's left of the anterior vagus nerve). A superficial incision (1–2 mm in depth) is made in the musculature of the distal 4–6 cm of the esophagus. The divided muscle is gently parted with a blunt hemostat until the underlying mucosa of the esophagus is encountered. The thickness of the muscle of the lower esophagus varies from a few millimeters to 0.5 cm or more. Great care must be taken to avoid opening into the lumen of the esophagus.



30.8a–c

The divided muscle is now separated from the underlying mucosa by blunt pledget dissection in the submucosal plane (**Figure 30.9a and b**). The dissection is continued until at least 50% of the circumference of the esophagus is free of the overlying muscle.



30.9a,b

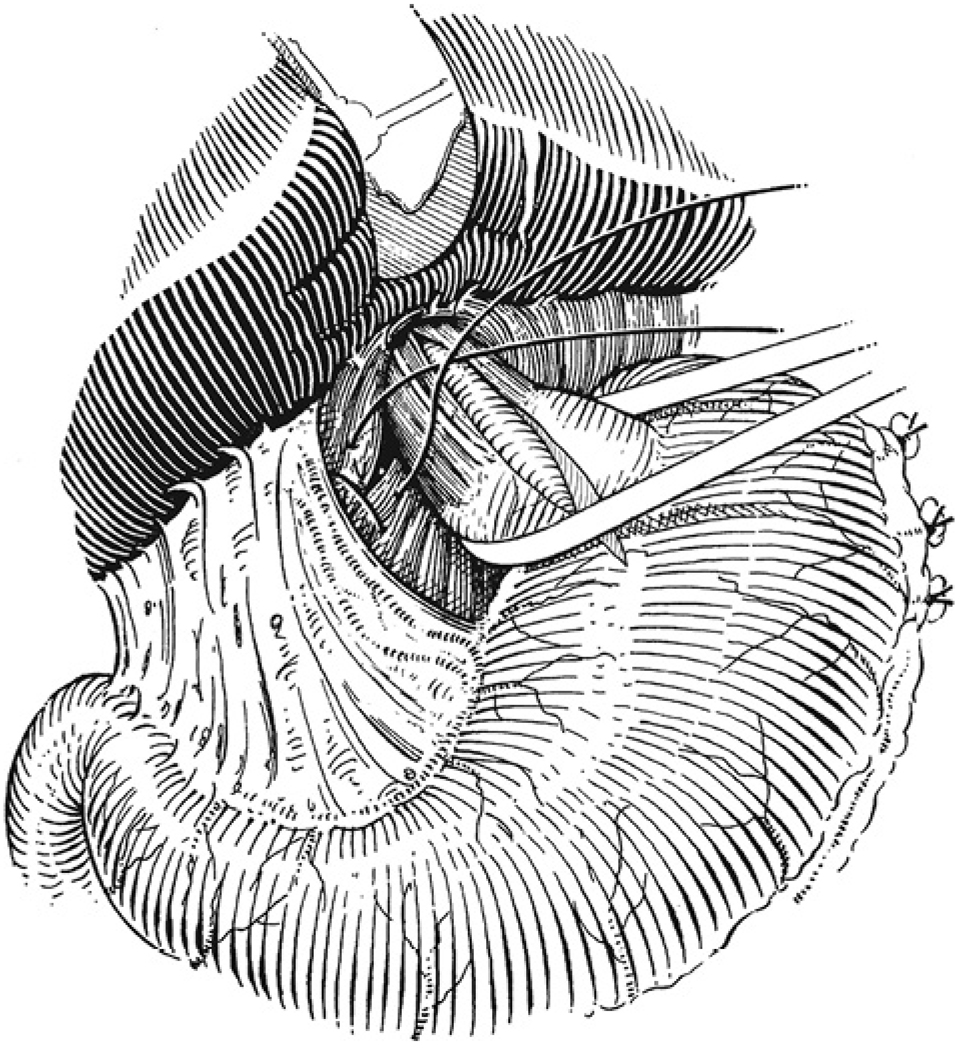
The myotomy is extended through the gastroesophageal junction for 1 cm onto the fundus of the stomach and the musculature is similarly elevated from the underlying mucosa.

TESTING FOR ESOPHAGEAL PERFORATION

The stomach and esophagus are distended with air introduced through the nasogastric tube, and the exposed mucosa is carefully inspected for perforation. A mucosal defect should be carefully closed with fine absorbable sutures.

NARROWING OF THE HIATUS

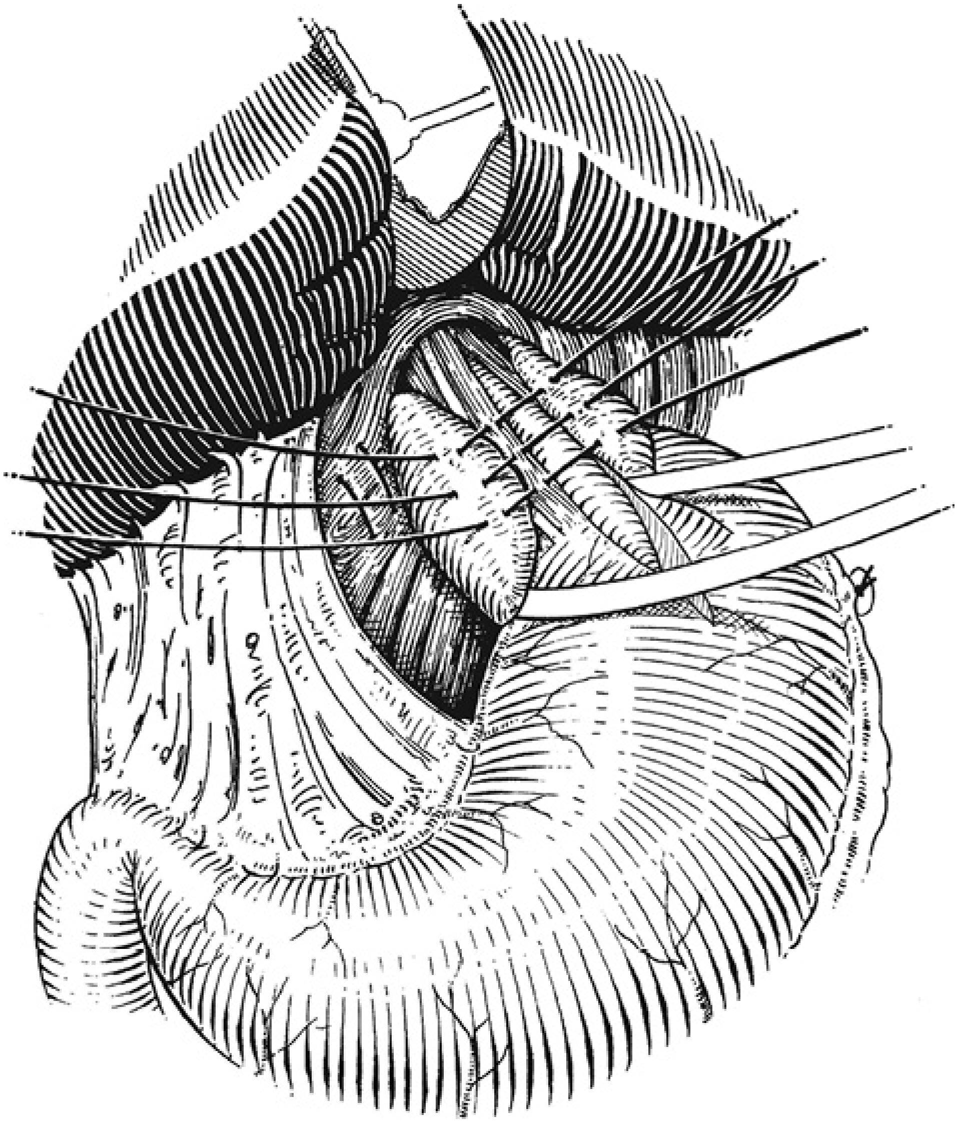
The esophageal hiatus is narrowed posteriorly to the esophagus by placing deep sutures through the crura of the diaphragm. The sutures are tied loosely to prevent them from cutting through, leaving sufficient space alongside the esophagus to allow passage of the tip of a finger. Two or three sutures may be required for this purpose (**Figure 30.10**).



30.10

FUNDOPLICATION

A loose (floppy) Nissen fundoplication is now constructed over the distal 1–1.5 cm of the esophagus. The esophageal sutures are only placed through one side of the divided esophageal muscle in order to prevent reapproximation of the edges of the myotomy (**Figure 30.11**).



30.11

Alternatively, an anterior Dor, or posterior Toupet fundoplication may be performed and attached to both aspects of the myotomy edges to keep the opposing edges from reapproximating.

WOUND CLOSURE

The wound is closed either in layers or with interrupted en masse sutures of 3/0 polyglycolic acid. A sub-cuticular suture approximates the skin edges.

Postoperative care

Nasogastric decompression and IV fluids are continued until the postoperative ileus has resolved (mean of 3–4 days).

Complications

Complications may include mediastinitis or peritonitis due to failure to detect a mucosal perforation, and recurrence of symptoms (dysphagia) if there is an incomplete myotomy. This may occur when the circular muscle is not separated from the underlying mucosa for at least half the circumference of the esophagus and/or the myotomy is not extended far enough onto the stomach. Gastroesophageal reflux may result from an inadequate fundoplication and dysphagia for solids may result from an overly tight fundoplication.

Outcome

After myotomy alone without an antireflux procedure, the long-term incidence of gastroesophageal reflux is around 15% but may be medically managed. Relief of the dysphagia and respiratory problems is usually complete, but residual or recurrent pain may occur in 25% of patients and is due to diffuse esophageal spasm. The esophageal pain generally responds to pneumatic dilatation.

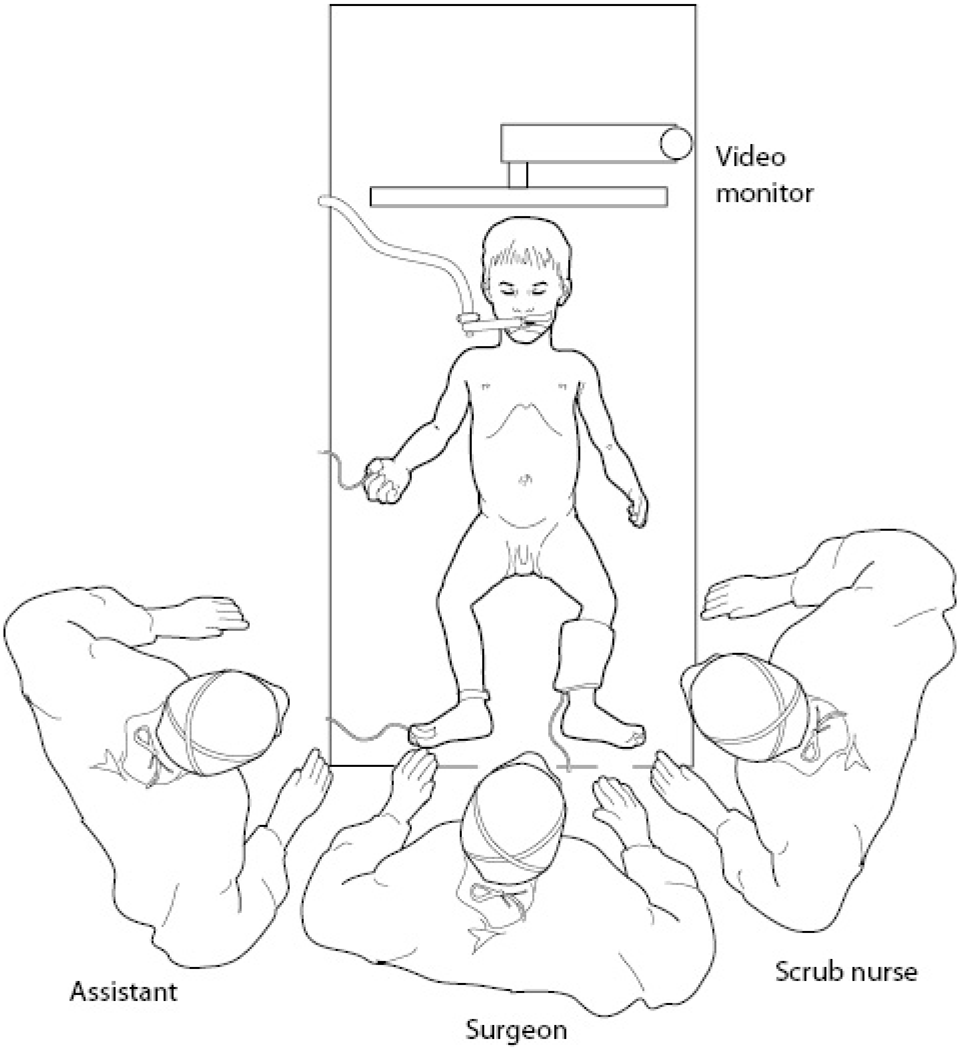
LAPAROSCOPIC MYOTOMY

Preoperative preparation and anesthesia

Neither bowel preparation nor placement of a Foley catheter into the patient's bladder is required before laparoscopic myotomy. General anesthesia with full relaxation and endotracheal intubation is mandatory. It is recommended to perform esophageal endoscopy and clear the esophagus from retained food after induction of anesthesia. Thereafter, a medium-size stiff nasogastric tube is inserted and left until the end of the operation to fixate the esophagus and to ensure that the stomach is empty for the duration of the operation. Single-shot IV perioperative antibiotic prophylaxis is given with induction of anesthesia.

PATIENT AND TEAM POSITIONING

The patient is placed in a supine position at the lower end of the operation table. The operation is performed by the surgeon who stands between the legs of the patient and the camera assistant is placed to his left. The scrub nurse is to the right hand of the surgeon. The laparoscopic tower including the video monitor is placed at the head or the left head of the table (**Figure 30.12**). The operation table is set in a reverse Trendelenburg position.



30.12

LAPAROSCOPIC INSTRUMENTATION

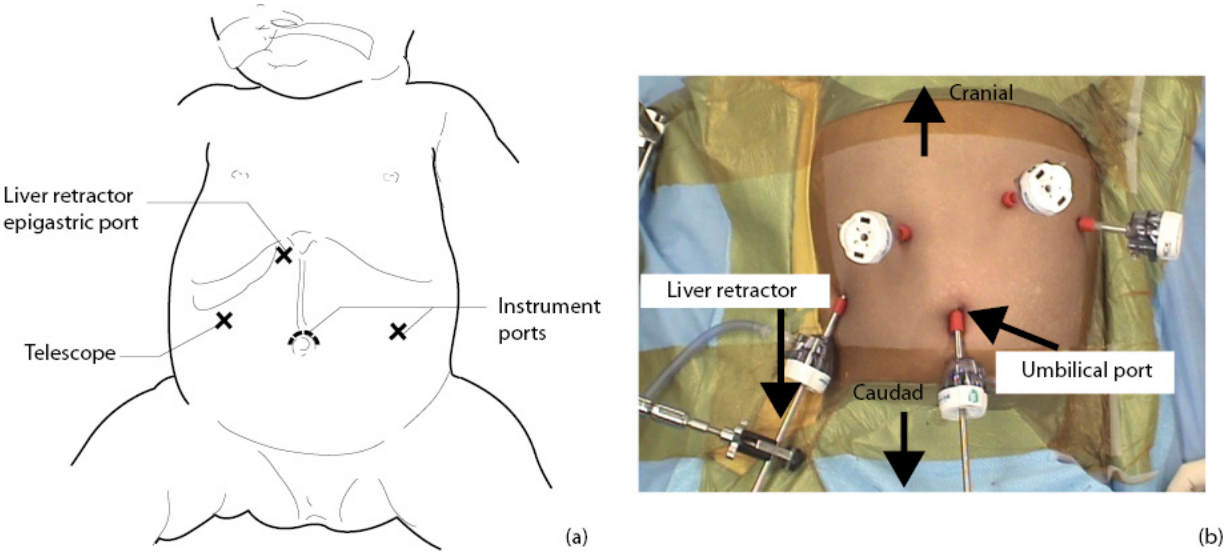
Most patients with symptomatic achalasia are older than 10 years of age. Therefore, 10 mm scopes and instruments with a diameter of 5 mm are used, however, the author prefers 3.5 mm ports in all children. The following instruments are required for laparoscopic Heller's cardiomyotomy:

- 30-degree telescope, 5 mm or 10 mm diameter
- 5 mm or 10 mm infraumbilical trocar and cannula
- 3.5 mm or 5 mm trocars and cannulae ×2 or 3
- Toothed, ratcheted grasper as liver retractor
- Curved scissors
- Tissue forceps ×2
- Needle holders ×2
- Monopolar hook
- Suction/irrigation device

Operation

PORT PLACEMENT

A four-port technique is used (**Figure 30.13a**). A 5 mm or 10 mm port for the telescope is inserted through or below the umbilicus with an open technique. Carbon dioxide is insufflated at a pressure of 8–10 mmHg. Two 3.5 mm or 5 mm instrument ports are inserted in the right and left mid-upper abdomen (one in each upper quadrant). A fourth trocar for the liver retractor may be introduced below the subcostal margin to the left of the falciform ligament. The grasper for liver retraction may also be introduced directly without using a trocar.

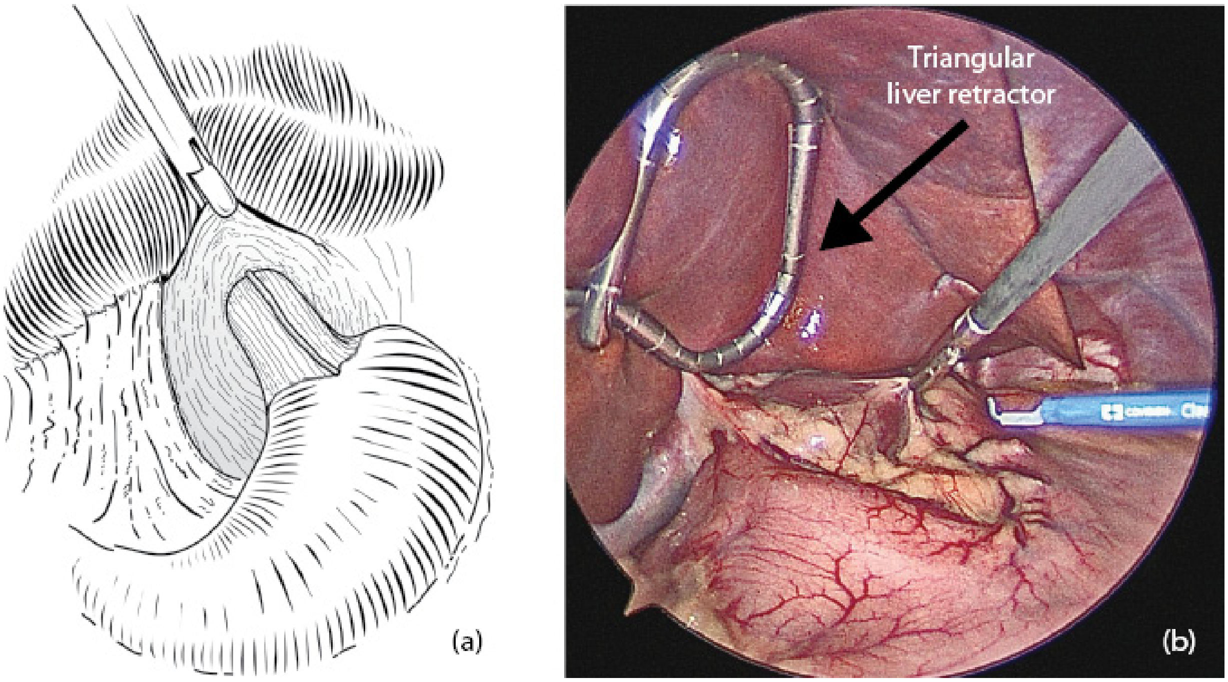

Liver retractor epigastric port
Telescope
Instrument ports
(a)
Cranial
Liver retractor
Umbilical port
Caudad
(b)

30.13a,b Port placement: (a) Four-port placement; (b) exterior view of the five-port technique.

In case of difficulties with exposure, an additional 5 mm port may be introduced above the umbilicus right to the falciform ligament for the telescope (**Figure 30.13b**). After changing the position of the telescope, an additional instrument may be used via the umbilical port for grasping the stomach and pulling it downward in these cases.

LIVER RETRACTION

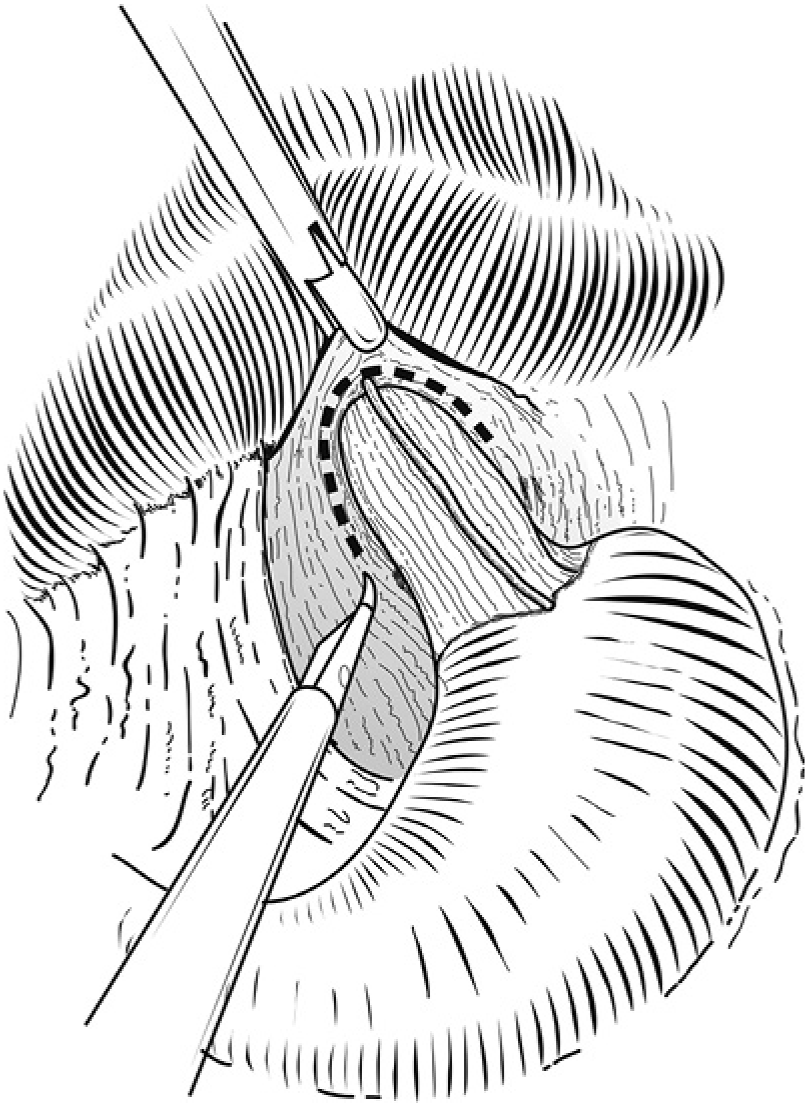
The left lobe of the liver can be easily retracted upward with a single-toothed ratcheted grasper, which is introduced through the subxiphoid incision with or without use of a port. The grasper is fixed to the muscular diaphragm by grasping it just above the hiatus (**Figure 30.14a**). It can then be left *in situ* for the duration of the operation. Alternatively, a triangular 5 mm liver retractor can be used to elevate the left lobe of the liver to expose the esophagogastric junction and hiatus (**Figure 30.14b**).



30.14a,b The left lobe of the liver can be retracted using a single-toothed ratcheted grasper (a) or a triangular 5 mm liver retractor (b).

EXPOSURE OF CARDIA AND ESOPHAGUS

The “white line,” which is the edge of the diaphragmatic crura, is easily identifiable by gentle downward traction of the stomach. The phrenoesophageal junction is divided using the monopolar hook and the anterior wall of the esophagus is freed (**Figure 30.15**). The dissection includes only the anterior and lateral esophagus; the posterior esophagus is not mobilized to prevent gastroesophageal reflux. The esophagus should be exposed from the crus down to the esophagogastric junction. The anterior vagal nerve should be identified, preserved, and pushed away from the myotomy incision.

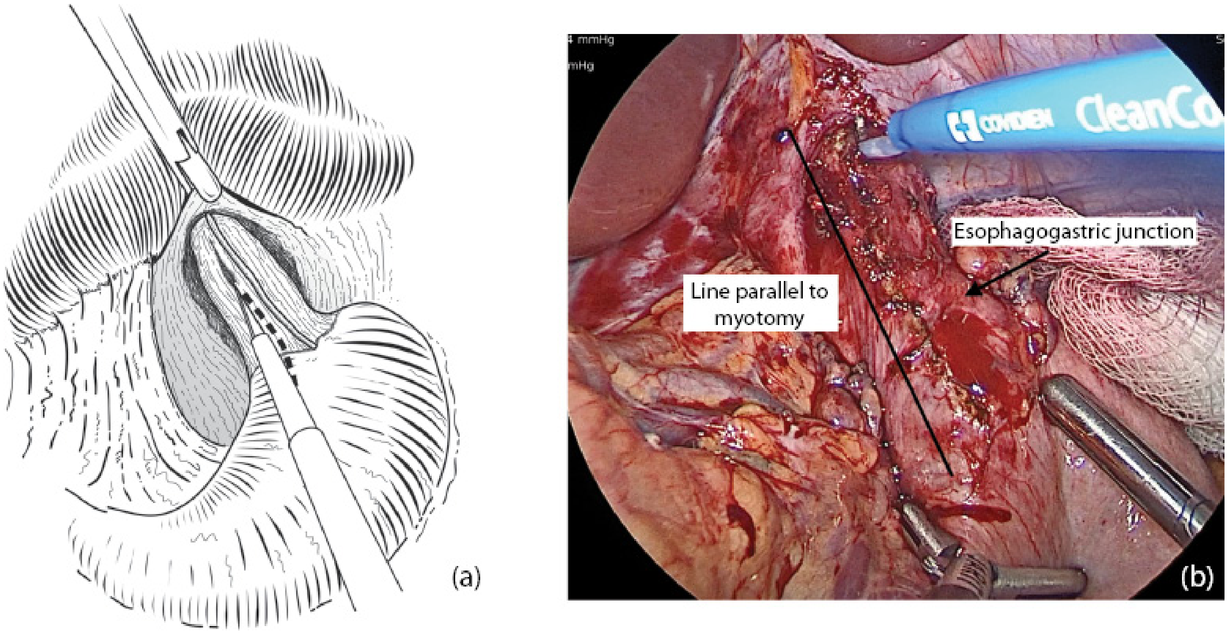


30.15 Division of the phrenoesophageal junction.

Thereafter, the anterior crus is lifted away from the esophagus to gain entry into the mediastinal esophagus. An easy plane can be developed between the overarching crus and the esophagus, allowing the esophagus to be exposed in the mediastinum for up to 5 cm. Once an adequate length of intrathoracic esophagus has been exposed, attention should be redirected to the abdominal esophagus.

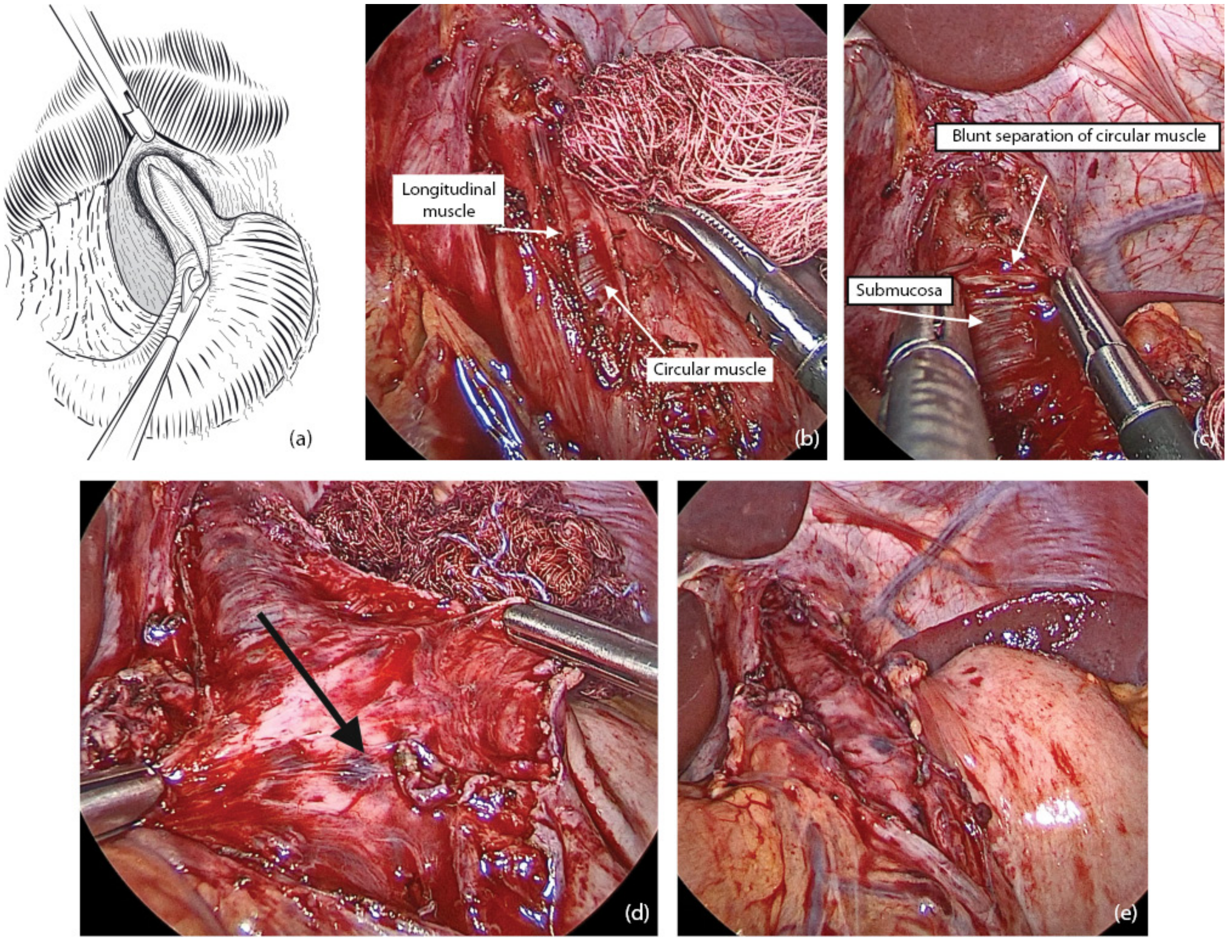
ESOPHAGEAL MYOTOMY

An esophageal myotomy (**Figure 30.16a and b**) is best started with a superficial incision on the anterior wall of the esophagus about 1 cm proximal to the esophagogastric junction. While it is possible to use monopolar diathermy or an ultrasonic scalpel to make this initial myotomy incision, both these instruments can produce deeper thermal damage, which may cause unrecognized damage of the underlying mucosa, resulting in delayed perforation.



30.16a,b (a) Esophageal myotomy. (b) Myotomy marked with cautery, the longitudinal muscle with split circular muscle beneath, and blunt separation of the circular muscles.

Blunt curved scissor dissection is used and the longitudinal muscle layers are separated (**Figure 30.17a**). The mucosa can be clearly seen as it herniates through the myotomy. The muscle layers should be spread further to the mediastinum for up to 5 cm using blunt dissection. Dissection is continued down to the gastric junction, extending onto the fundus of the stomach, which can be identified when the edges of circular gastric muscle fibers are seen (**Figure 30.17b–d**). The underlying mucosa should be allowed to pout outwards as much as possible. Bleeding from the esophageal muscle layers usually stops spontaneously and cauterization is unnecessary.


(a)
Longitudinal muscle
Circular muscle
(b)
Blunt separation of circular muscle
Submucosa
(c)
(d)
(e)

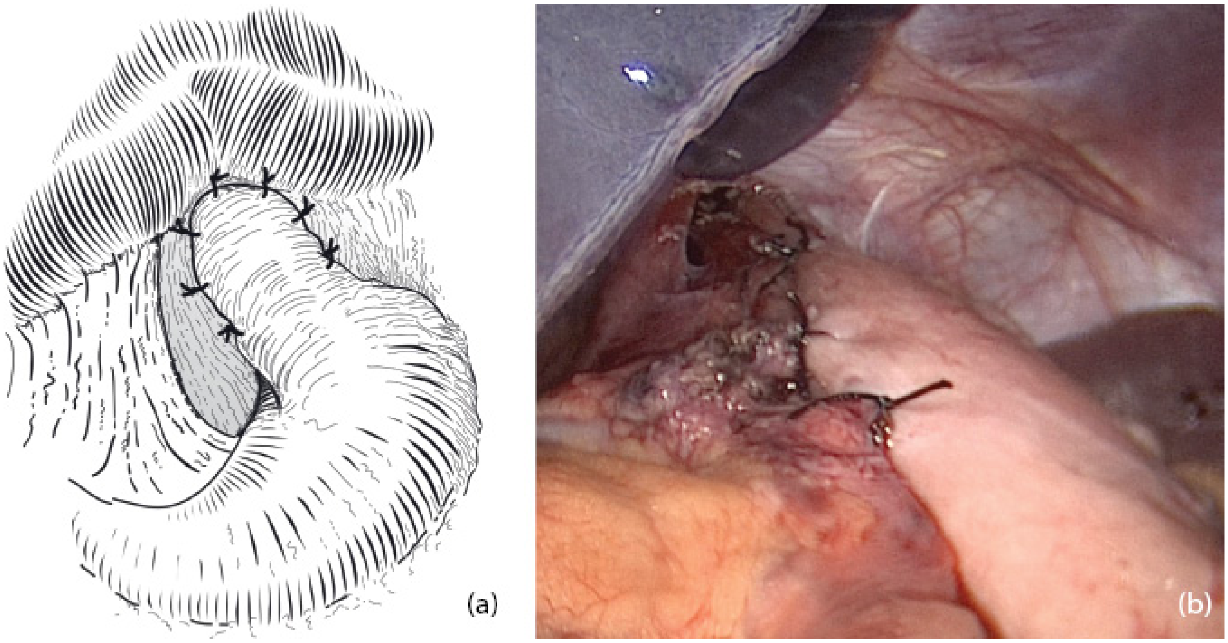
30.17a–e (a) Blunt curved scissor dissection is used to separate the longitudinal muscle layers. (b) The longitudinal muscle is split and circular muscle is visible beneath. (c) Blunt separation of the circular muscle with submucosal evident beneath. (d) Extension of myotomy onto the stomach. (e) Intraoperative view of the completed myotomy.

The mucosa is inspected for evidence of perforation and, if there is concern, air may be instilled into the esophagus and stomach to check for mucosal leak. A mucosal leak can be repaired laparoscopically with an absorbable mucosal suture.

Intraoperative endoscopy may facilitate identification of the esophagogastric junction (Figure 30.17e) and dynamic manometry or EndoFLIP balloon measurements may identify the adequate length of myotomy, but these maneuvers are optional.

FUNDOPLICATION

A 180-degree anterior fundoplication is performed to prevent reflux and to protect the mucosa (**Figure 30.18a and b**). The anterior gastric fundus is fixed to the anterior esophageal muscle layers with between four and seven non-absorbable sutures. The upper sutures are attached to the anterior hiatus to prevent slipping of the wrap. There is no need to divide the short gastric vessels.



30.18a,b Anterior Dor fundoplication.

Postoperative care

The nasogastric tube is removed immediately after anesthesia. Patients may usually commence feeding with liquids immediately following the operation, with immediate relief of symptoms. A routine contrast study is not required before commencement of feeding. Oral feeding should be completed by postoperative day 3.

Long-term follow-up is essential and clinical assessment is performed at 1, 6, and 12 months postoperatively. Contrast study, pH study, and manometry should only be used in case of persistent or new symptoms.

Complications

Bleeding from the myotomy site is common and usually stops spontaneously without further treatment.

Perforation of the mucosa during myotomy is rare and occurs mainly at the esophagogastric junction. Absorbable sutures are used to close the perforation site. Patients with missed or delayed perforation present with sepsis and symptoms of peritonitis or mediastinitis within 24–48 hours postoperatively. A contrast study is required to confirm perforation, and conventional surgical therapy via laparotomy or laparoscopy is required. As with fundoplication, there is a small risk of breaching the left pleural cavity during dissection of the anterior wall of the mediastinal esophagus, and a tension pneumothorax may require immediate drainage.

Long-term dysphagia may occasionally require balloon dilatation or repeat myotomy to deal with an incomplete myotomy or a too-tight fundoplication.

PERORAL ENDOSCOPIC MYOTOMY

History

The clinical application of peroral endoscopic myotomy (POEM) for achalasia was first reported by Inoue in 2010. This technique utilizes a completely endoscopic method for performing the esophagocardiomyotomy. Adoption of this method has been rapid in adults with achalasia and increasing numbers of pediatric patients are undergoing POEM for the condition.

Preoperative preparation and anesthesia

Patients should consume a liquid-only diet for 5 days prior to surgery to prevent accumulation of food debris within the esophagus. After IV antibiotic administration and endotracheal intubation, initial flexible endoscopy is performed to ensure the esophagus is clear.

PATIENT AND TEAM POSITIONING

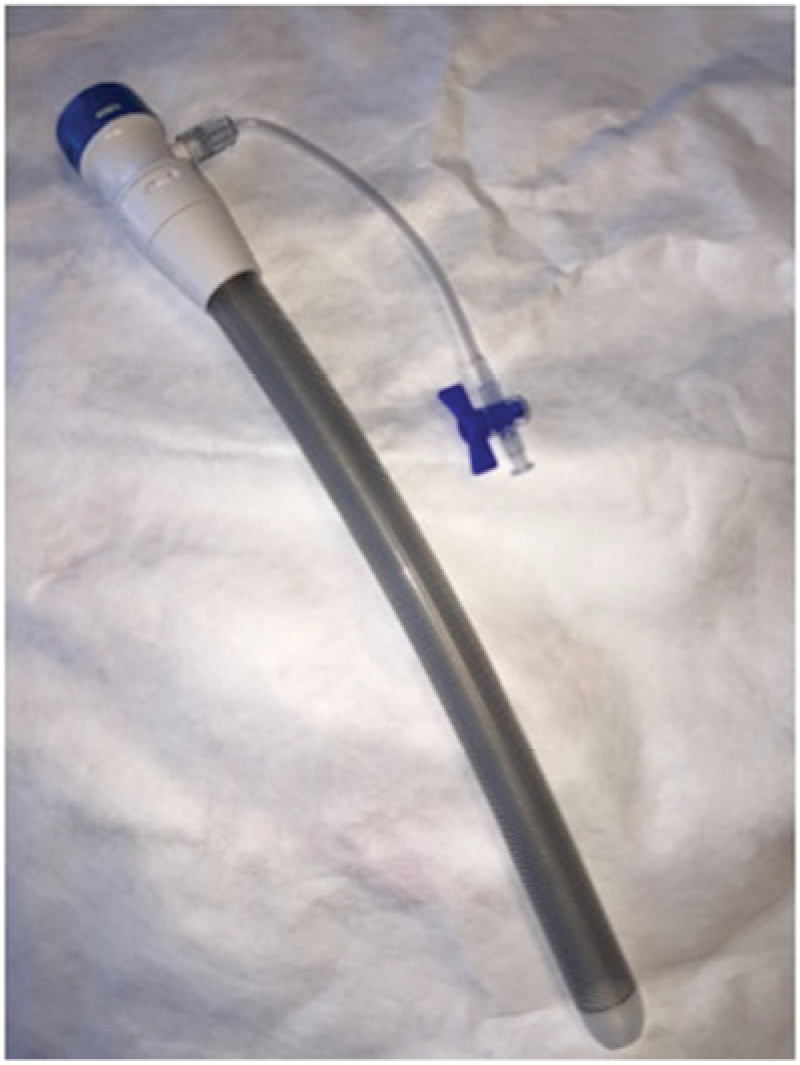
With the patient in a supine position, the surgeon stands at the head of the table and the assistant to the patient's left side. The surgical technician is either at the foot of the table or at the patient's right side. The abdomen should be exposed so that abdominal distension from pneumoperitoneum may be monitored and evacuated if necessary.

INSTRUMENTATION

- Video tower; Olympus GIF-H180 endoscope with CO_2 insufflation
- Lifting solution (2 mL methylene blue in 100 mL normal saline)
- Injection needle: Olympus NM-400L-0423
- Needle knife: Boston Scientific M00545840; triangle-tip knife: Olympus KD-640L
- Dissection caps: Hard Olympus MH-588; angled Olympus MAJ-Y0173
- Hemostatic graspers: Olympus FD-411UR or LR
- Apollo OverTube™ (Apollo Endosurgery)
- ERBE VIO 300D generator and ground pad (ERBE USA)
- Clips: Large Boston Scientific Resolution clip M0052260 (open/close); large Cook Instinct clip G18343 (Cook Medical)

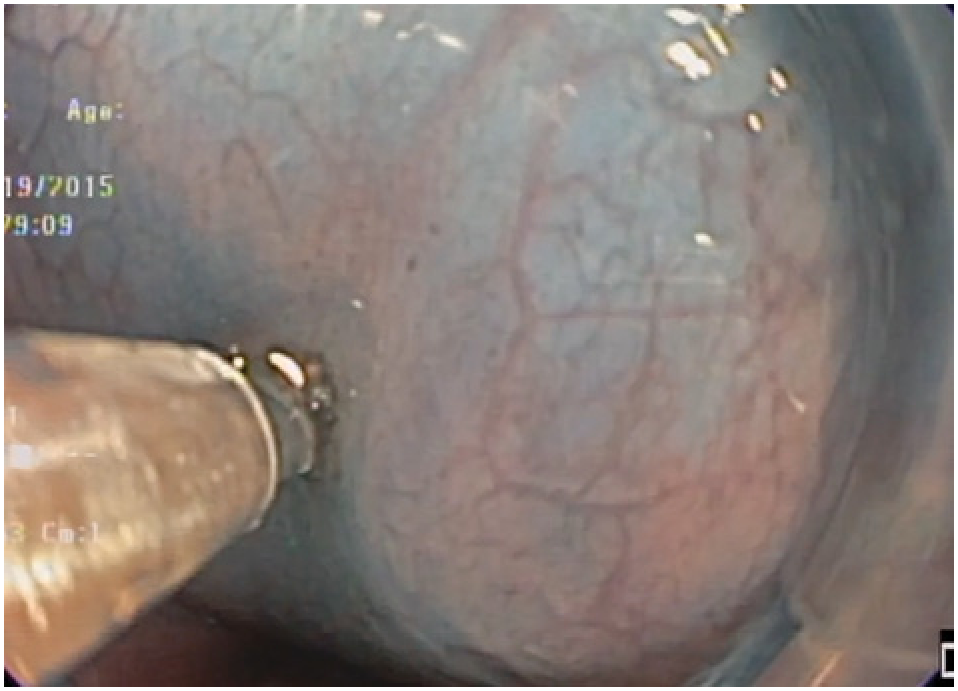
Operation

Endoscopy is performed and important measurements are taken to identify the esophagogastric junction (EGJ), mucosotomy site (about 10–15 cm proximal to the EGJ at the 2 o'clock position), the myotomy start and endpoints, and other landmarks. Measurements can be confirmed using an OverTube (Apollo Endosurgery, Austin, TX, USA) (**Figure 30.19**).



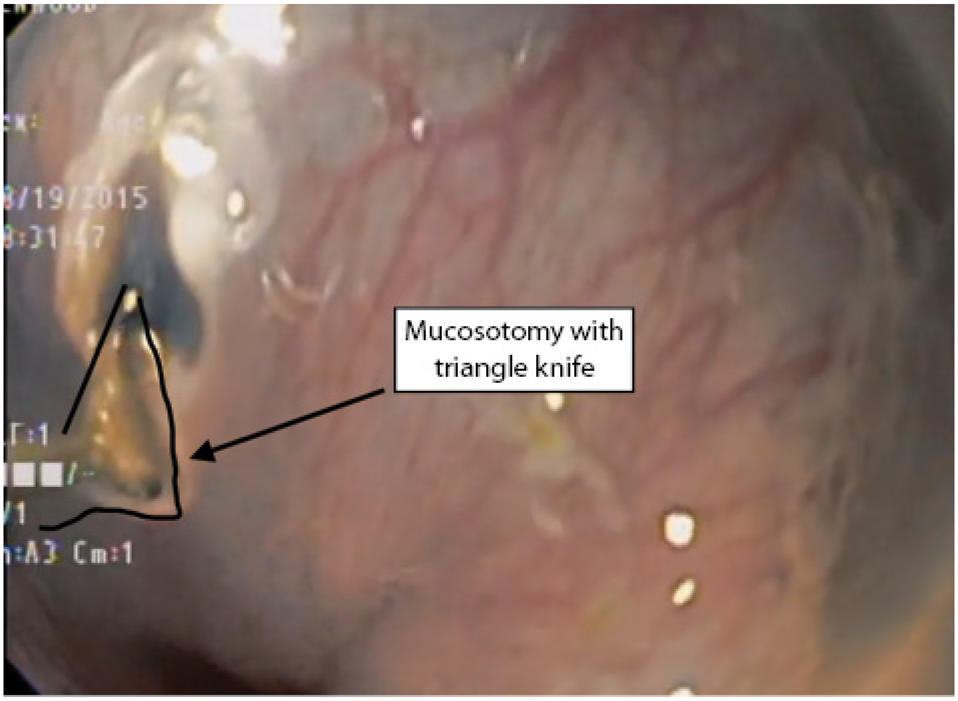
30.19 The Apollo OverTube.

Five milliliters of methylene blue solution is injected into the submucosal space at the site of the mucosotomy (2 o'clock position) (**Figure 30.20**).



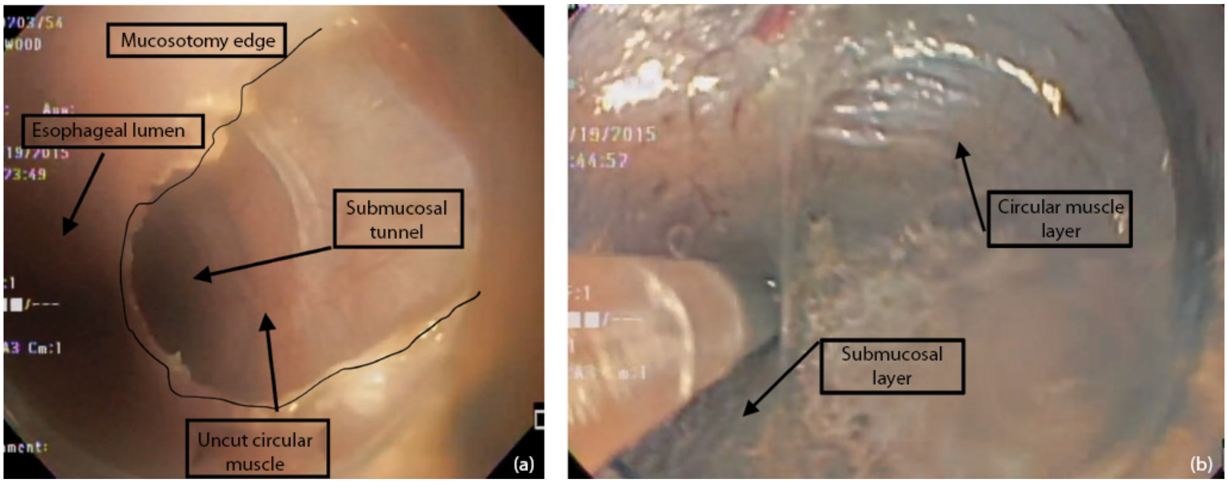
30.20 Methylene blue solution is injected into the submucosal space.

The needle knife or triangle knife is used to incise the mucosa for a distance of 2 cm (large enough to accept the dissecting cap) (**Figure 30.21**). The ERBE settings used are 60 Endocut and effect 4.



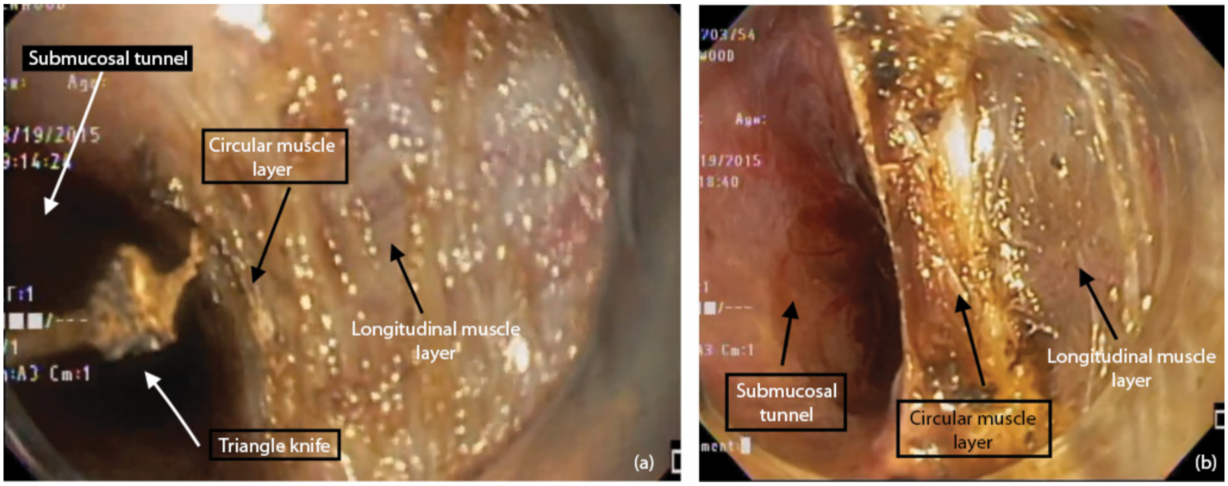
30.21 Incising the mucosa.

The endoscope with dissecting cap is insinuated into the layer between the submucosal and muscular layers. A triangle knife, needle knife, or L-hook may be used to create the tunnel down past the EGJ (**Figure 30.22a and b**) with the ERBE cautery settings on 60 spray and effect 2. The endoscope is removed from the tunnel and traversed through the main esophagus to ensure an adequate submucosal tunnel on the gastric aspect.


Mucosotomy edge
Esophageal lumen
Submucosal tunnel
Uncut circular muscle
(a)
Circular muscle layer
Submucosal layer
(b)

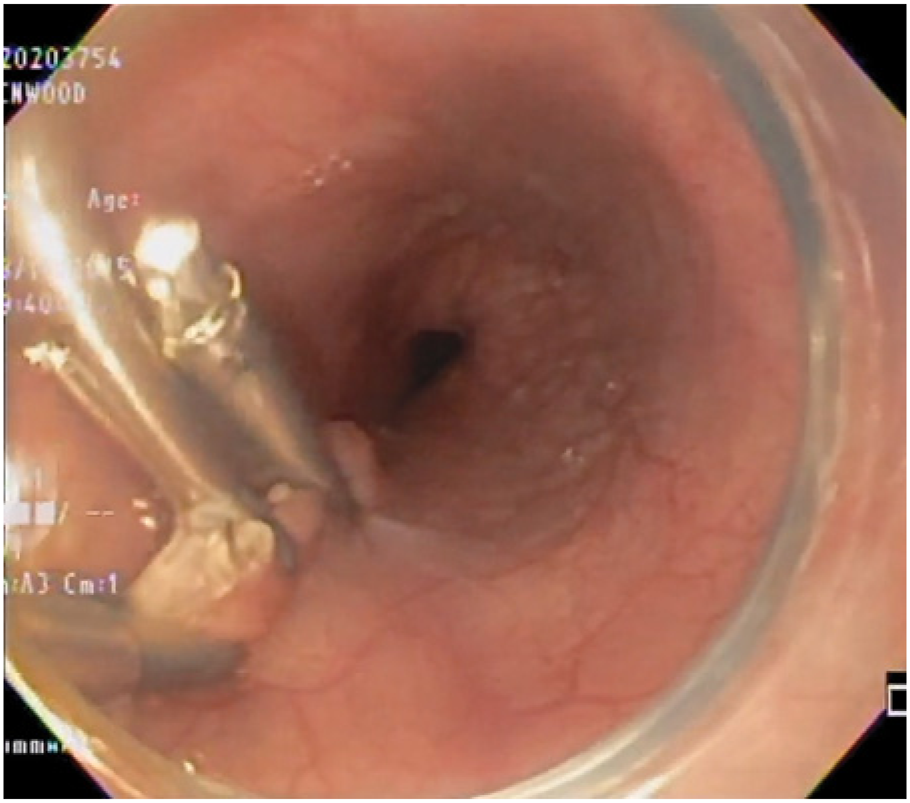
30.22a,b Submucosal tunnel shown (a) proximally and (b) distally.

The myotomy is performed with ERBE cautery of 40 spray and effect 2 for a length (as individualized per patient) beginning proximally in the esophagus and extending 2–3 cm onto the gastric side (**Figure 30.23a and b**).


Submucosal tunnel
Circular muscle layer
Longitudinal muscle layer
Triangle knife
(a)
Submucosal tunnel
Circular muscle layer
Longitudinal muscle layer
(b)

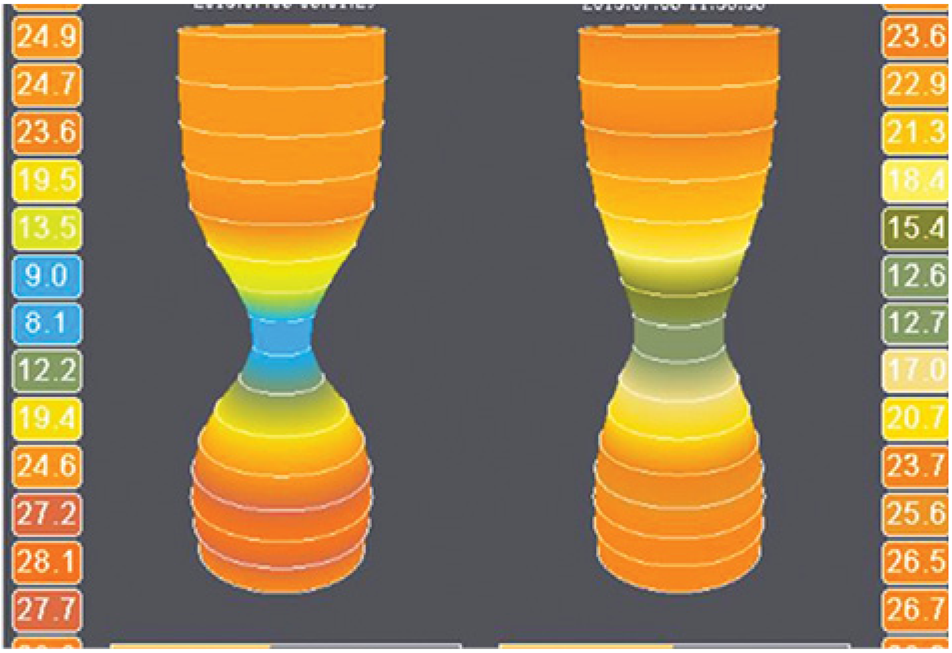
30.23a,b The myotomy is performed with ERBE cautery, beginning proximally in the esophagus and extending onto the gastric side.

Once the myotomy is completed, the mucosotomy defect is most commonly closed with Resolution™ clips (Boston Scientific, Marlborough, MA, USA) (**Figure 30.24**).



30.24 The mucosotomy is closed with Resolution clips.

More complicated suturing devices have been used (Apollo OverStitch™; Apollo Endosurgery Inc.; Austin, TX, USA) but require a dual lumen endoscope to perform. The endoscopic functional lumen imaging probe (EndoFLIP®; Medtronics, Minneapolis, MN, USA) may be used during both laparoscopic Heller myotomy and POEM procedures to aid in identifying the adequacy of myotomy using cross-sectional diameter measurements of the EGJ before and after myotomy, as described by Teitelbaum et al. (**Figure 30.25**).



30.25 In this case, the cross-sectional diameter of the EGJ has been improved from 8.1 mm to 12.6 mm following POEM.

Postoperative care

Following POEM, patients are kept nil per os overnight and undergo an esophagram on postoperative day one. This is performed to rule out an esophageal leak, assess the patency of the EGJ, and serve as a baseline study in order to compare to subsequent studies in the future if necessary. Patients are started on soft diet immediately after esophagram and instructed to continue for the first 2 weeks after POEM. Similar to laparoscopic Heller myotomy, long-term follow-up is required at 1, 6, and 12 months following POEM in order to monitor for persistent or recurrent symptoms.

Complications

Bleeding in the submucosal tunnel may occur but is usually managed endoscopically. Full-thickness perforation of the esophageal or gastric wall during POEM may lead to pneumoperitoneum, pneumothorax, or pneumomediastinum. If clinically significant, drainage is performed by laparoscopic trocar or Veress needle, or tube thoracostomy, respectively. Rarely, operative repair for significant gastric perforation during POEM (drain placement or laparoscopic or open repair) has been described. Gastroesophageal reflux may be more common with POEM since a fundoplication is not performed, but this is usually manageable with medication. Similarly, in patients with recurrent or persistent dysphagia, incomplete myotomy should be considered and managed accordingly with dilation or repeat myotomy.

VIDEOS

OPS_8e_video_30.1 Laparoscopic Heller
OPS_8e_video_30.2 POEM

FURTHER READING

Donahue PE, Samelson S, Schlesinger PK, et al. Achalasia of the esophagus: Treatment, controversies and the method of choice. *Ann Surg* 1986; 203: 505–11.

Emblem R, Stringer MD, Hall CM, Spitz L. Current results of surgery for achalasia of the cardia. *Arch Dis Child* 1993; 68: 749–51.

Inoue H, Minami H, Kobayashi Y, et al. Peroral endoscopic myotomy (POEM) for esophageal achalasia. *Endoscopy* 2010; 42: 265–71.

Inoue H, Sato H, Ikeda H, et al. Per-oral endoscopic myotomy: A series of 500 patients. *J Am Coll Surg* 2015; 221: 256–64.

Mattioli G, Prato AP, Castagnetti M, Jasonni V. Esophageal achalasia. In: Bax K(N)MA, Georgeson KE, Rothenberg SS, et al. (eds). *Endoscopic Surgery in Children*. Berlin: Springer; 2008: pp. 245–52.

Franklin AL, Petrosyan M, Kane TD. Childhood achalasia: A comprehensive review of disease, diagnosis and therapeutic management. *World J Gastrointest Endosc* 2014; 6(4): 105–11.

Petrosyan M, Khalafallah AM, Guzzetta PC, et al. Surgical management of esophageal achalasia: Evolution of an institutional approach to minimally invasive repair. *J Pediatr Surg* 2016; 51(10): 1619–22.

Smits M, van Lennep M, Vrijlandt R, et al. Pediatric achalasia in the Netherlands: Incidence, clinical course, and quality of life. *J Pediatr* 2016; 169: 110–15.

Teitelbaum EN, Soper NJ, Pandolfino JE, et al. Esophagogastric junction distensibility measurements during Heller myotomy and POEM for achalasia predict postoperative symptomatic outcomes. *Surg Endosc* 2015; 29: 522–8.

Zaninotto G, Annese V, Costantini M, et al. Randomized controlled trial of botulinum toxin versus laparoscopic Heller myotomy for esophageal achalasia. *Ann Surg* 2004; 239: 364–70.

31

Gastrostomy

ALEJANDRA M. CASAR BERAZALUCE, AARON P. GARRISON, AND TODD A. PONSKY

HISTORY

Gastrostomy is one of the oldest abdominal operations in continuous use, and its history is closely associated with the evolution of modern surgery.

Although pediatric surgeons have become more selective in the use of gastrostomies for congenital malformations, there has been a marked increase in their use in children without associated surgical pathology, mainly those with inability to swallow secondary to central nervous system (CNS) disorders. Refinements in traditional procedures and the introduction of newer and simpler endoscopically, radiologically, and laparoscopically aided gastrostomies have enhanced the safety and expanded the applicability of this operation. It is fair to state that few procedures have had as many different approaches as this seemingly simple access to the stomach. The use of softer, minimally irritating materials in the manufacture of gastrostomy catheters and the availability of several types of skin-level gastrostomy devices, commonly referred to as "buttons," have greatly facilitated the long-term use of this type of enterostomy.

PRINCIPLES AND JUSTIFICATION

Gastrostomy is indicated in infants and children for long-term feeding, decompression, or a combination of both. It is also commonly employed in conjunction with other interventions, like antireflux procedures. Additional uses of gastrostomy include gastric access for esophageal bougienage, placement of transpyloric jejunal feeding tubes, and administration of medications.

Indications

There is no real consensus on the timing and indications for gastrostomy, as many indications for supplemental feeding via g-tube have been shown to resolve or improve after early infancy.

The type of gastrostomy, preoperative workup, technique, and choice of gastrostomy device depend mainly on the reason for the procedure, the child's age and underlying disease, and the familiarity of the surgeon with the different surgical techniques.

Placement of a gastrostomy for enteral feeding has two prerequisites:

- The upper gastrointestinal tract must be functional.
- The need for enteral feedings must be long-term (at least several months).

Children benefiting from gastrostomy fall into two broad categories: Those who are unable to swallow, and those unable to consume adequate nutrients orally. The first group is larger, and is composed primarily of patients with neurologic disturbances, oropharyngeal disorders, and oral aversion. The second group includes patients with a variety of conditions in which the CNS is intact: Failure to thrive, complex bowel disorders (e.g. short gut syndrome, Crohn's disease, malabsorption), malignancy and other debilitating illnesses, and various congenital or acquired diseases interfering with growth.

In selected patients, a gastrostomy is the most effective means of administering a non-palatable special diet (e.g. chronic renal failure) or ensuring compliance with medication (e.g. administering cholestyramine in Alagille's syndrome, antiretroviral therapy in HIV).

Because of the high incidence of foregut dysmotility in neurologically impaired patients, there has been an increase in the use of jejunal feeding through combined gastrostomy–jejunostomy or skin-level devices. Alternatively, jejunostomies are, at times, added to gastrostomy procedures.

When a gastrostomy is placed in conjunction with another intra-abdominal procedure (such as fundoplication or repair of duodenal atresia), the tube is used initially for decompression and then for intragastric feedings. If prolonged gastric or duodenal dysmotility is anticipated, a smaller, more flexible catheter can be advanced into the lumen of the jejunum, exiting either along the gastrostomy tube (gastrojejunostomy) or as a jejunal feeding tube through a separate incision. Gastrostomies can be a temporary adjunct in the management of children with severe pathologic aerophagia and be of palliative value in the management of patients with intestinal obstruction secondary to unresectable malignancy. Most gastrostomy types are suitable for these purposes.

Caveats and contraindications

With all gastrostomy techniques, great care must be exercised in children with previous epigastric procedures, hepato- and/or splenomegaly, and in patients with severe musculoskeletal abnormalities, such as scoliosis. For the percutaneous technique, contraindications are the inability to perform upper tract endoscopy safely or to identify transabdominal illumination and see an anterior gastric wall indentation clearly. Anatomic abnormalities (such as malrotation or marked scoliosis), ascites, coagulopathy, and severe intra-abdominal infection may render the procedure inadvisable. In such cases, laparoscopic or radiologic control, a fully laparoscopic procedure, or conventional laparotomy may be indicated.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

In general, patients who can tolerate bolus orogastric or nasogastric feeds are good candidates for gastrostomy placement alone. Caution should be taken when placing one in patients with failure to thrive, gastroesophageal reflux, or respiratory difficulties associated with feeding, as they may not tolerate gastric feeds. A trial of nasogastric feeds can help predict success of gastrostomy in this population. If nasogastric feedings are not tolerated, which usually manifests with vomiting or an inability to feed the stomach in less than 30–60 minutes, patients may be better served by nasojejunal/gastrojejunal access or fundoplication.

Before taking a patient to the operating room, the surgeon should be sure that there is no need for simultaneous fundoplication; if any question exists, a reflux workup should be completed. The precise workup for gastroesophageal reflux is beyond the scope of this chapter and varies widely among institutions. Some surgeons routinely perform an upper gastrointestinal series in all patients undergoing gastrostomy to evaluate for malrotation; however, its routine use has been studied by multiple groups and found to be of extremely low yield.

Gastroesophageal reflux is a common problem in neurologically impaired children both before and after the placement of a gastrostomy. Preoperative evaluation of the degree of reflux in these patients is necessary. Patients with severe reflux are best managed with an antireflux procedure in addition to a gastrostomy. Children with mild or no reflux are candidates for gastrostomy only.

The addition of a gastrostomy to the surgical correction of a congenital or acquired condition should be considered only if it will substantially facilitate perioperative or long-term care. This decision should be made preoperatively, when possible, to ensure adequate surgical planning. Examples in neonatal surgery include complex esophageal atresia, certain duodenal obstructions, select abdominal wall defects in which long-term ileus is anticipated, and short gut syndrome. Indications in older children include severe esophageal stricture, complex foregut trauma, intestinal pseudo-obstruction, malignancy, and complex adhesive bowel obstruction.

OPERATIONS

A variety of options exists for placement of gastrostomy tubes including open (mainly Stamm), endoscopic (PEG), laparoscopic, hybrid, and percutaneous image-guided approaches.

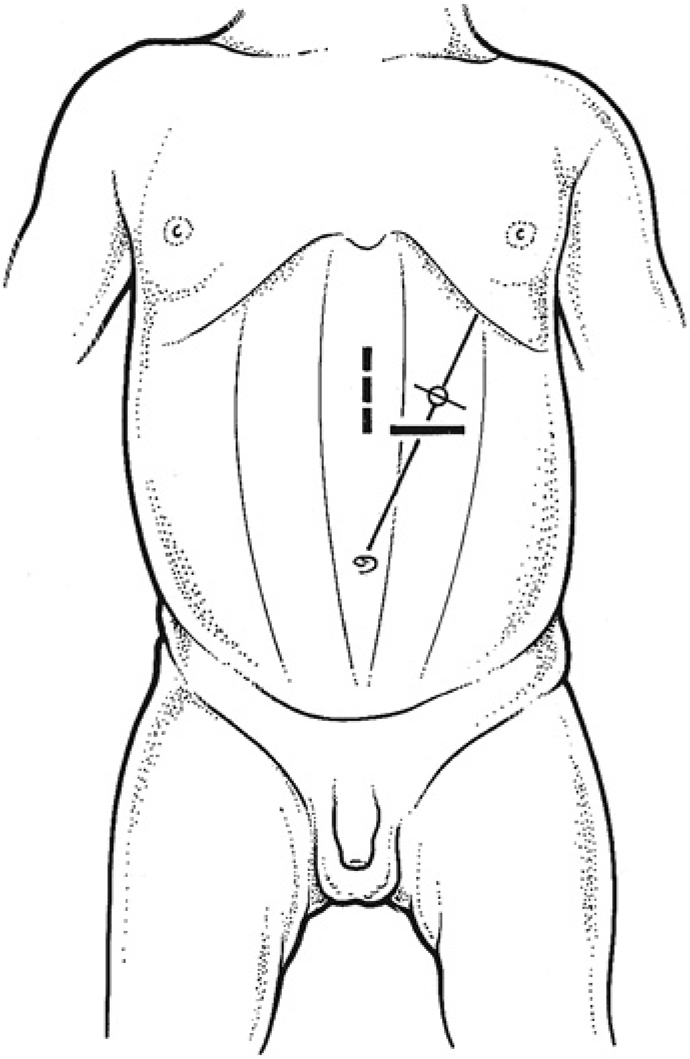
Open technique

STAMM GASTROSTOMY

The open Stamm technique was first devised in the 1890s and involved an upper midline laparotomy with a catheter brought out through a separate incision in the left upper quadrant.

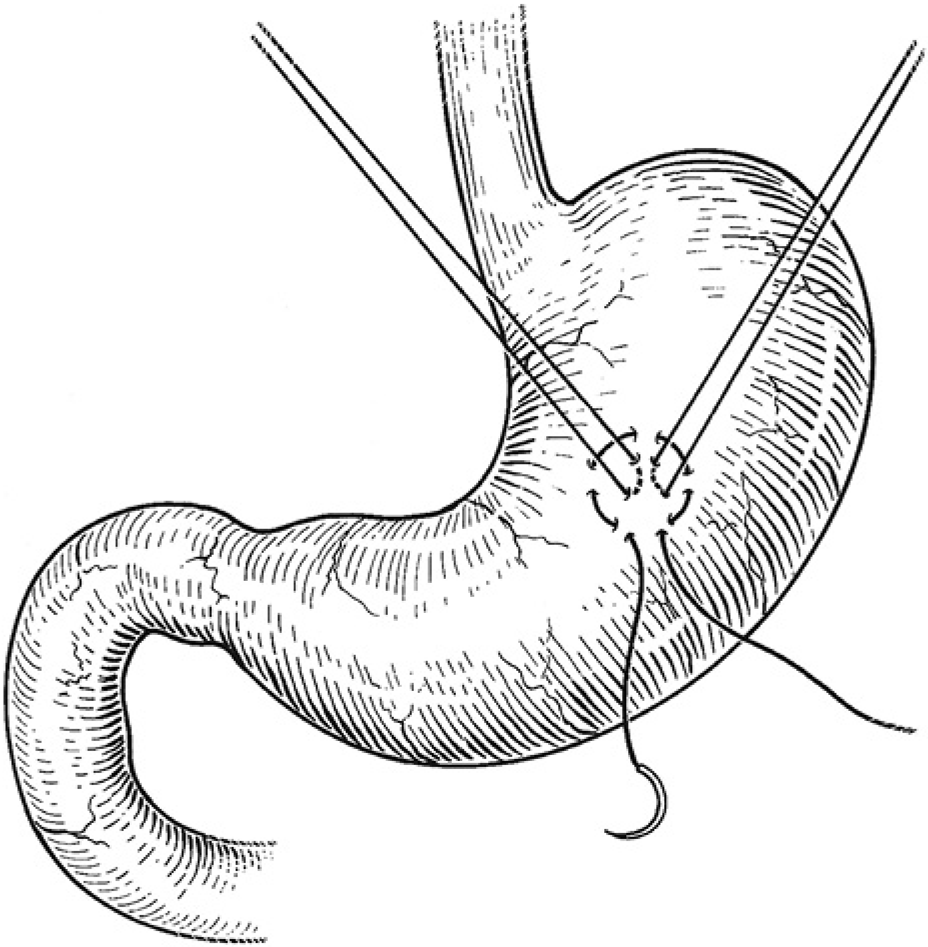
The child is positioned with a small roll behind the back to elevate the epigastrium, then prepared and draped. We use a primary button gastrostomy ranging in size from 12 Fr to 14 Fr in full-term neonates to 20 Fr for adolescents, and a 10 Fr T-tube or Malecot catheter for preterm infants or neonates with very small stomachs, such as those found in children with esophageal atresia without distal tracheoesophageal fistula.

For pediatric patients, the stomach is approached through a short, left, transverse, supraumbilical incision (**Figure 31.1**). Fascial layers are incised transversely, and the muscle is retracted or transected. The catheter exit site is approximately at the junction of the lower two-thirds and the upper one-third of a line from the umbilicus to the mid-portion of the left ribcage, over the mid-rectus muscle. A vertical incision may be useful in children with a high-lying stomach or a narrow costal angle.



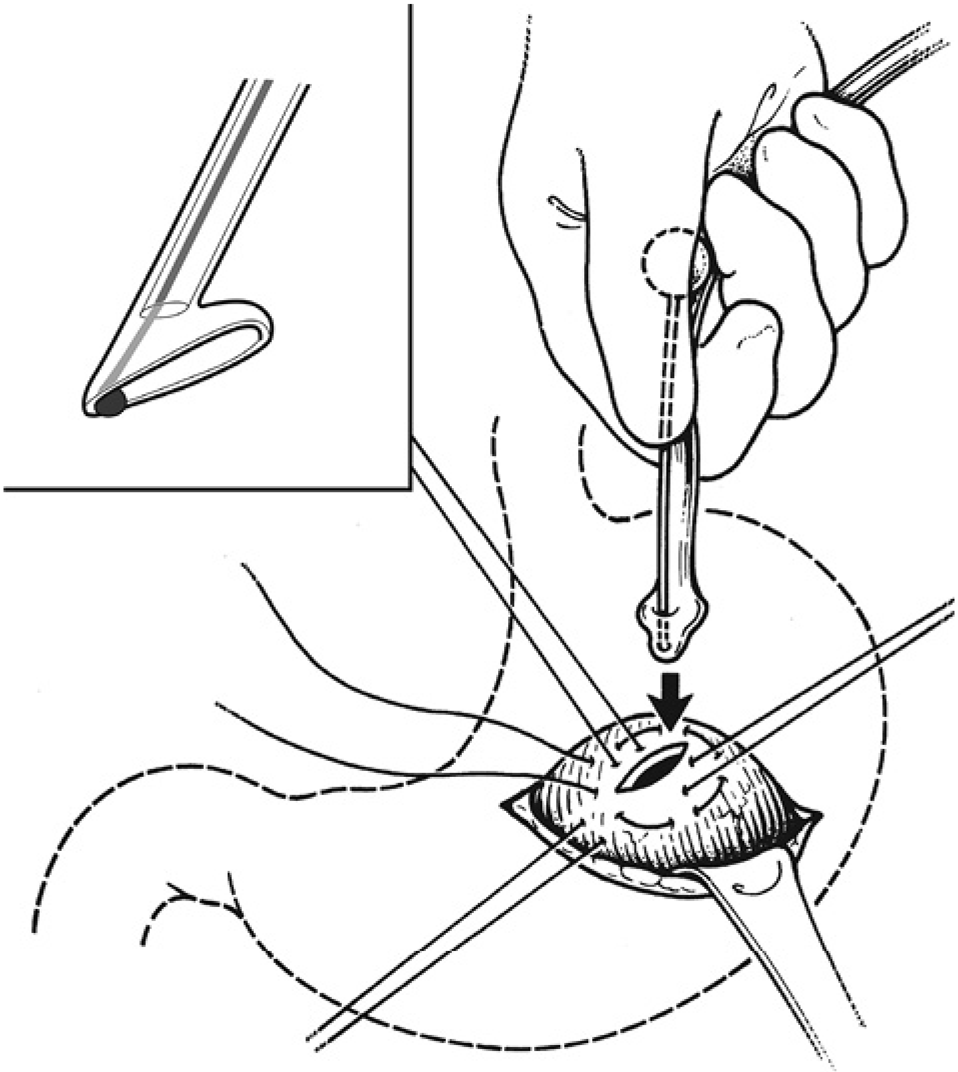
31.1

Traction sutures and a purse-string suture (synthetic, absorbable) are placed as shown in **Figure 31.2**. The opening should be away from the gastric pacemaker at the level of the splenic hilum; away from the greater curvature because that site may be needed for construction of a gastric tube for esophageal replacement; away from the fundus to allow for a possible fundoplication; and away from the antrum to prevent excessive leakage and pyloric obstruction by the catheter tip. If the catheter is to be placed cranially and close to the lesser curvature for a gastrostomy with antireflux properties, care must be taken to avoid the vagus nerve.



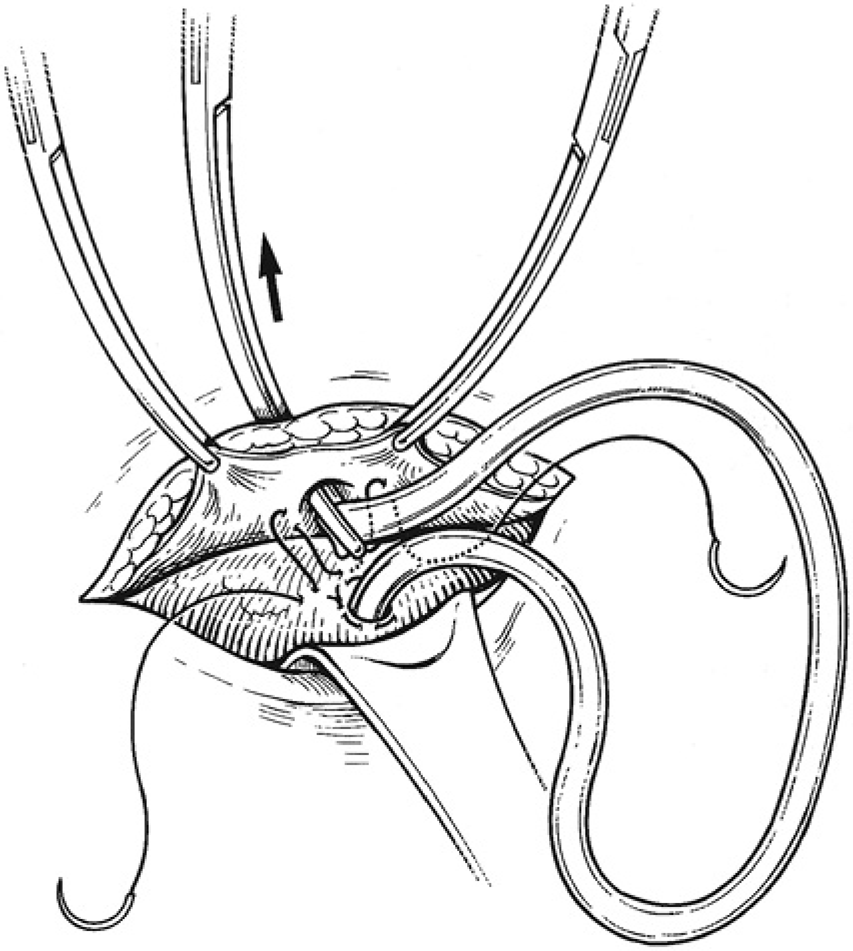
31.2

A lower tacking suture pulls the stomach caudally, enhancing exposure and allowing better gastric access. The gastrotomy is performed with fine scissors or cautery, while the upper sutures are lifted to prevent injuring the back wall. The catheter is introduced using a simple stylet while these sutures are elevated (**Figure 31.3**).



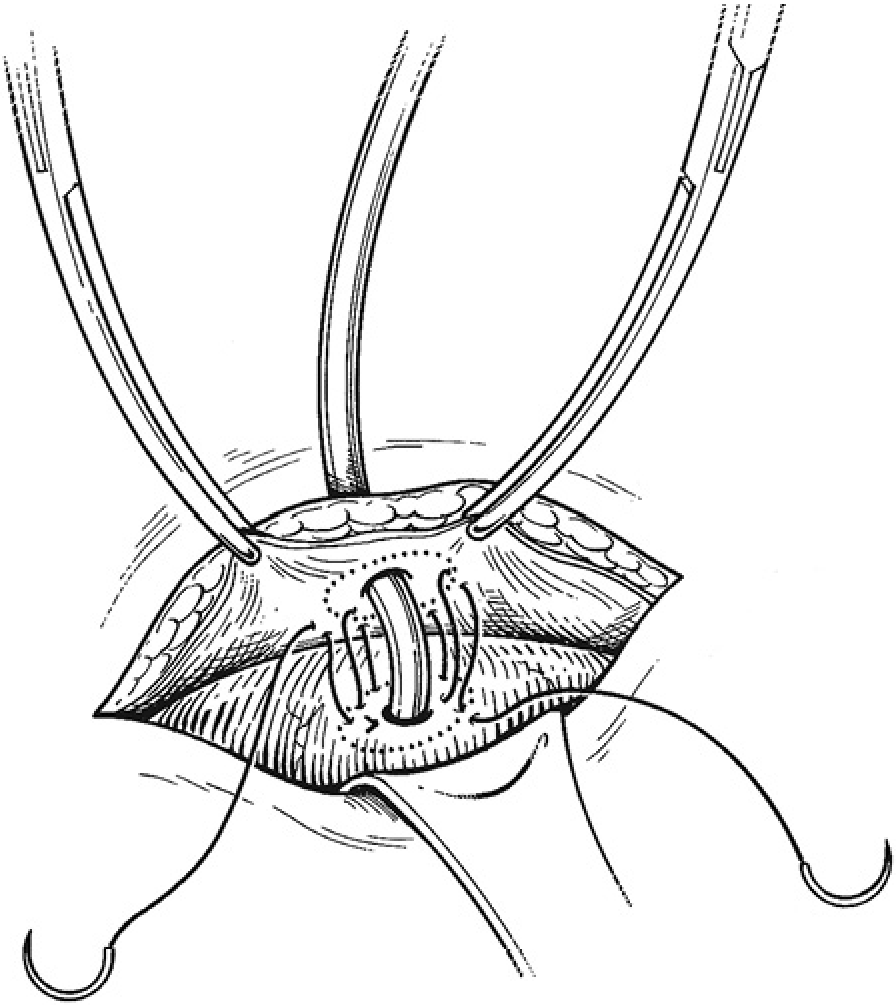
31.3

The purse-string suture is tied. A continuous, synthetic, absorbable, monofilament suture (polydioxanone) is used to anchor the stomach to the anterior abdominal wall. A Kelly clamp is placed through a counter-incision and the abdominal wall layers are pushed inward. The posterior 180 degrees of the anastomosis are completed, the peritoneum and fascia are incised, and the tip of the clamp is pushed through. The catheter end is grasped, and the tube is brought out through the counter-incision (**Figure 31.4**).



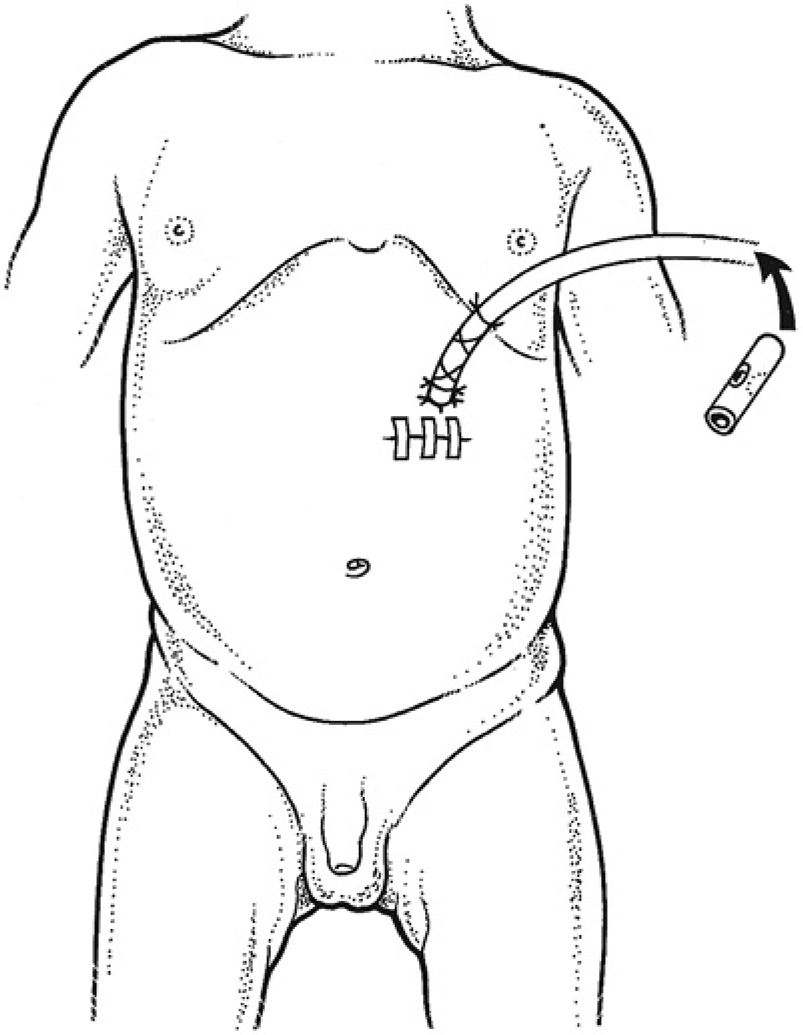
31.4

Placement of the continuous monofilament suture is now completed (**Figure 31.5**). When tied, this suture provides a 360-degree fixation with a watertight seal. In most cases, this maneuver obviates the need for a second purse-string suture.



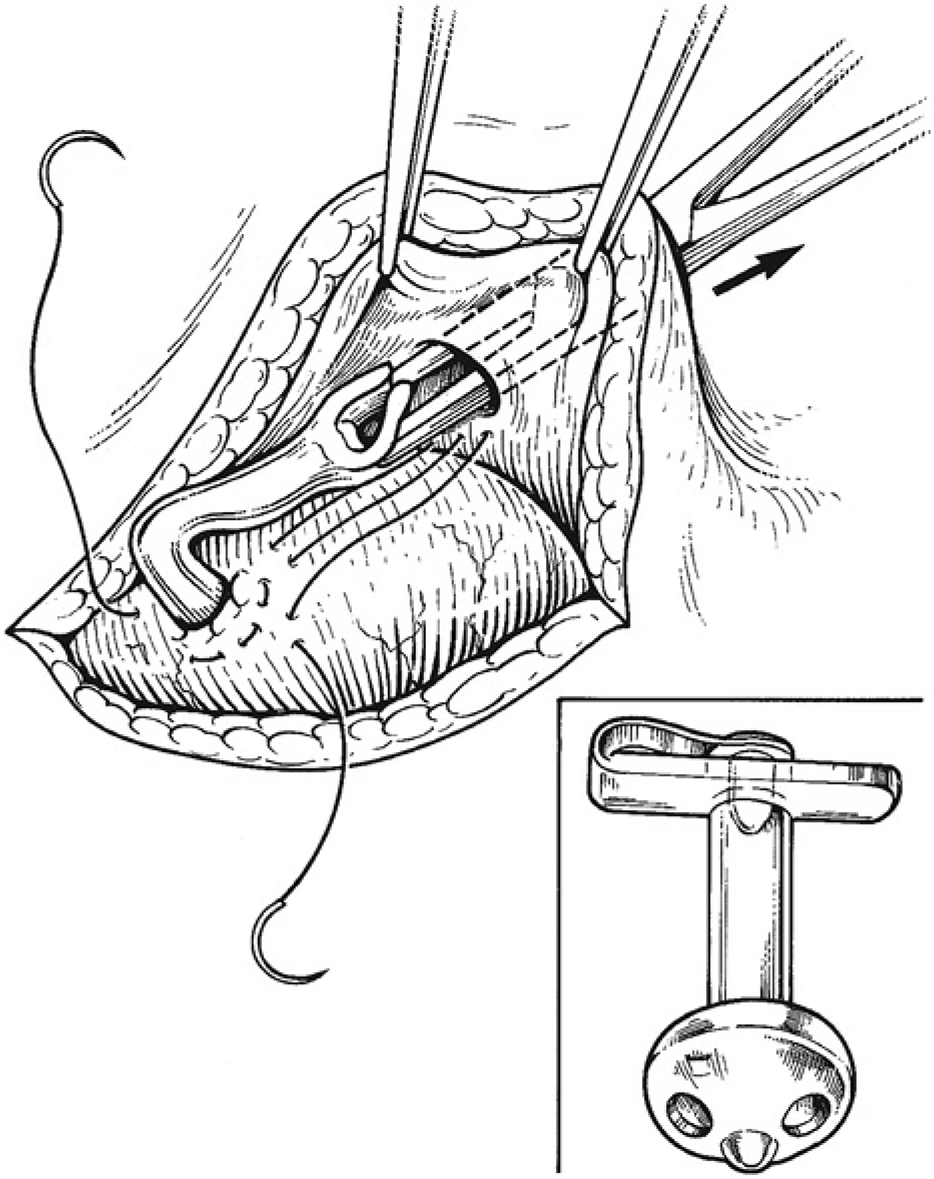
31.5

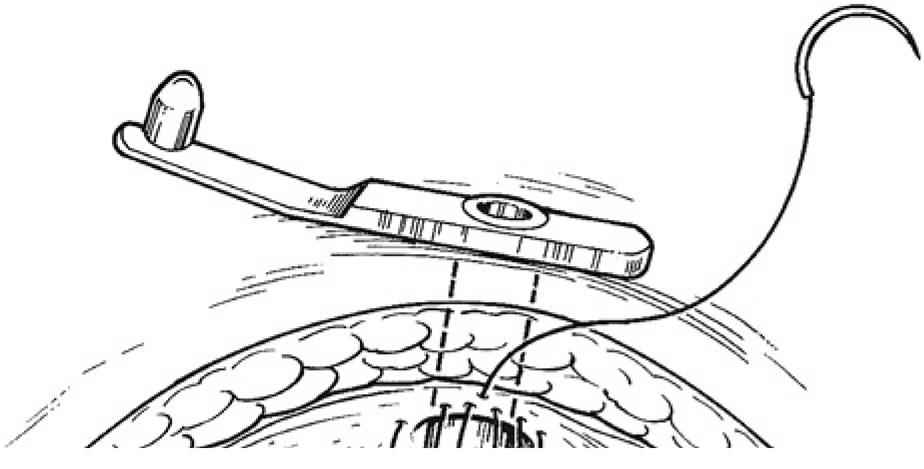
The abdominal wall layers are closed with synthetic absorbable sutures and the skin is approximated with subcuticular stitches and adhesive strips. The catheter is secured with synthetic monofilament sutures (polypropylene). These are removed 1–2 weeks after the operation, and a small crossbar is placed to prevent distal catheter migration (**Figure 31.6**).

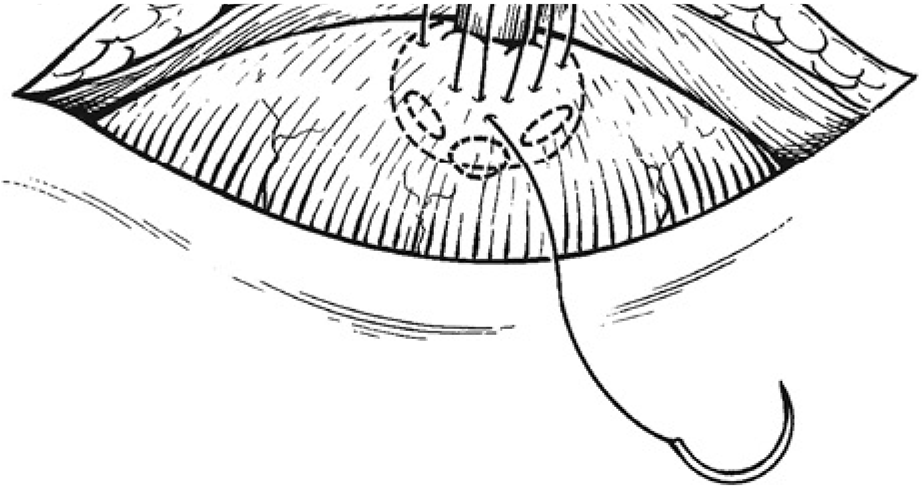


31.6

This standard procedure may be modified to allow the insertion of other devices, available in different shaft lengths and diameters (**Figure 31.7**). The shaft length should encompass the invaginated gastric wall, the abdominal wall, and an additional few millimeters of "play" to allow for postoperative edema, ease of care, and subsequent growth and weight gain.

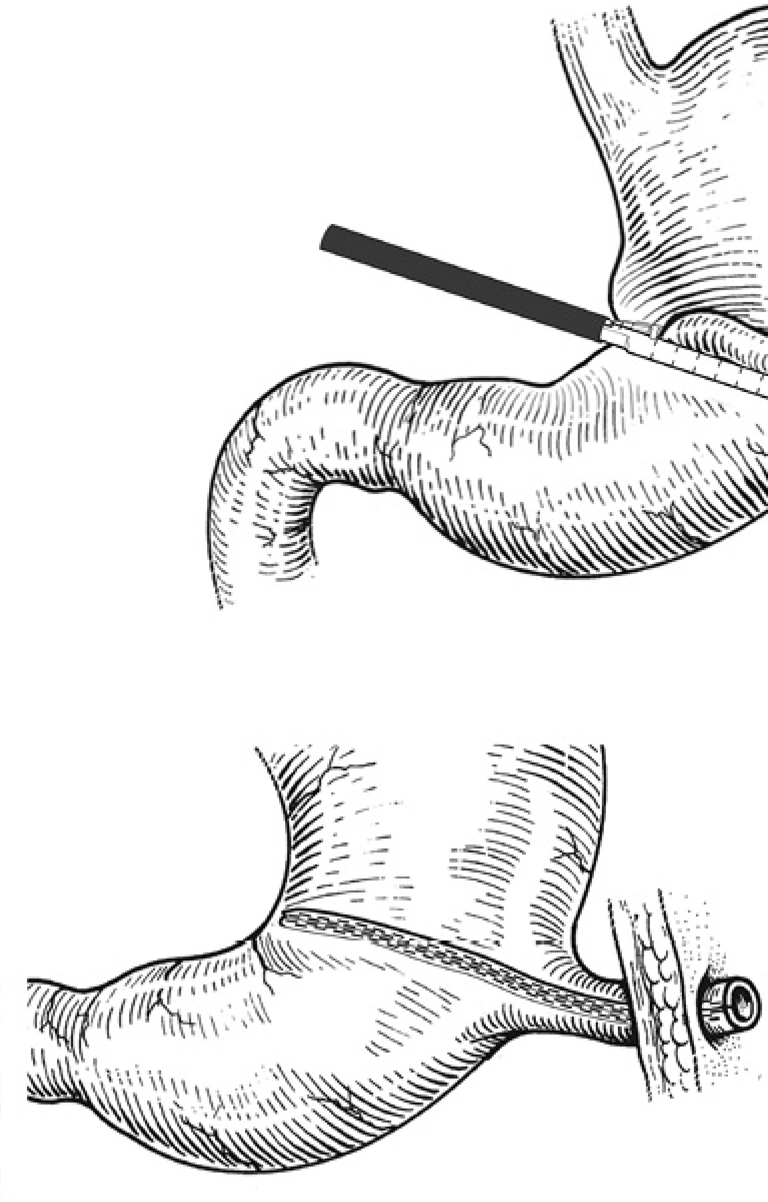






31.7

The stomach is vented with a nasogastric tube or a special decompression device which, when inserted in the shaft, deactivates the one-way valve at the gastric end (**Figure 31.8**).



31.8

JANEWAY GASTROSTOMY

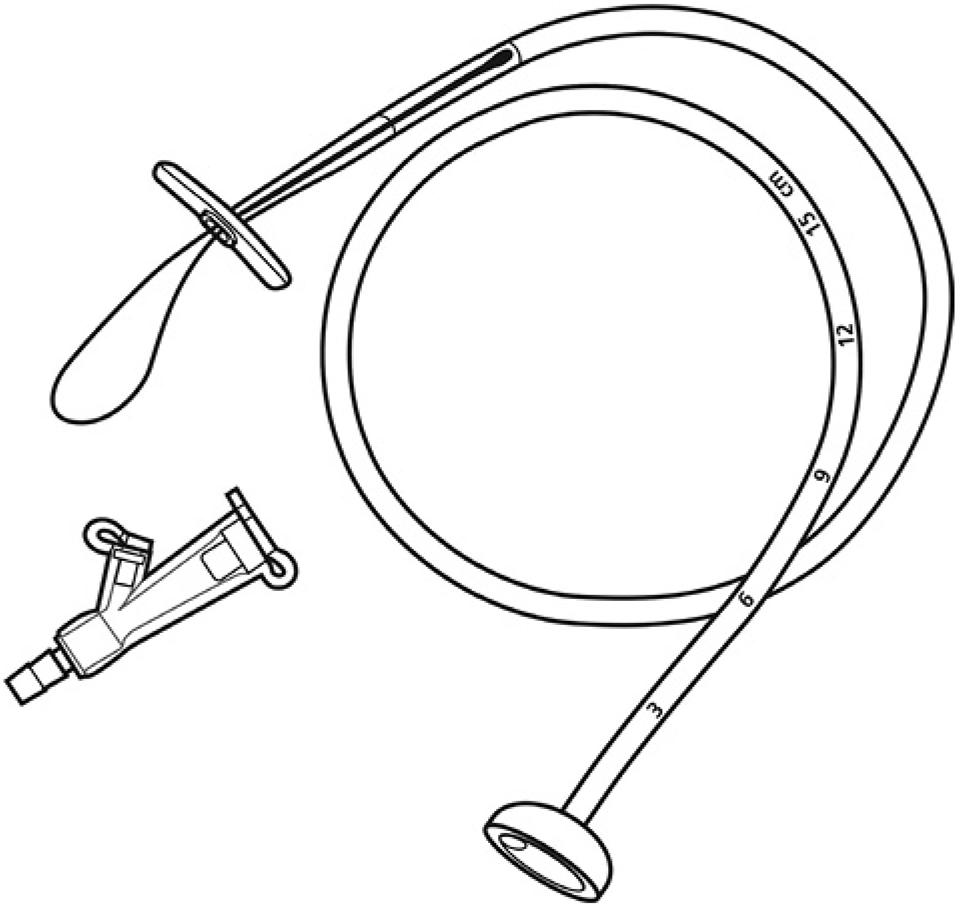
This procedure may be accomplished either using a conventional laparotomy with a gastrointestinal anastomosis stapler or employing an endoscopic stapler under laparoscopic control. The stapler is employed to tubularize the gastric wall (**Figure 31.8**). The gastric tube is brought out away from the incision (if open) or through one of the port sites (if laparoscopic).

Percutaneous endoscopic gastrostomy ("pull" technique)

The percutaneous endoscopic gastrostomy (PEG) technique was first performed in pediatrics in 1979 by Gauderer and Ponsky. The procedure is best performed in the operating room. Older children and those able to tolerate endoscopy without compromising the upper airway can receive local anesthesia with sedation as needed. Younger children require general endotracheal anesthesia, primarily because of anticipated difficulties with airway management. A single dose of a broad-spectrum IV antibiotic is given shortly before the procedure. For endoscopy, the smallest available pediatric gastroscope is used. A 16 Fr silicone rubber catheter is suitable for most children. Hybrid catheters for primary implantation of skin-level devices are also available.

A note of caution: In children with marked anatomical distortions, such as severe scoliosis, the stomach may be displaced cranially into the left chest. The transverse colon is then pulled upward and is more vulnerable to puncture. In these cases, laparoscopic control adds safety to the procedure.

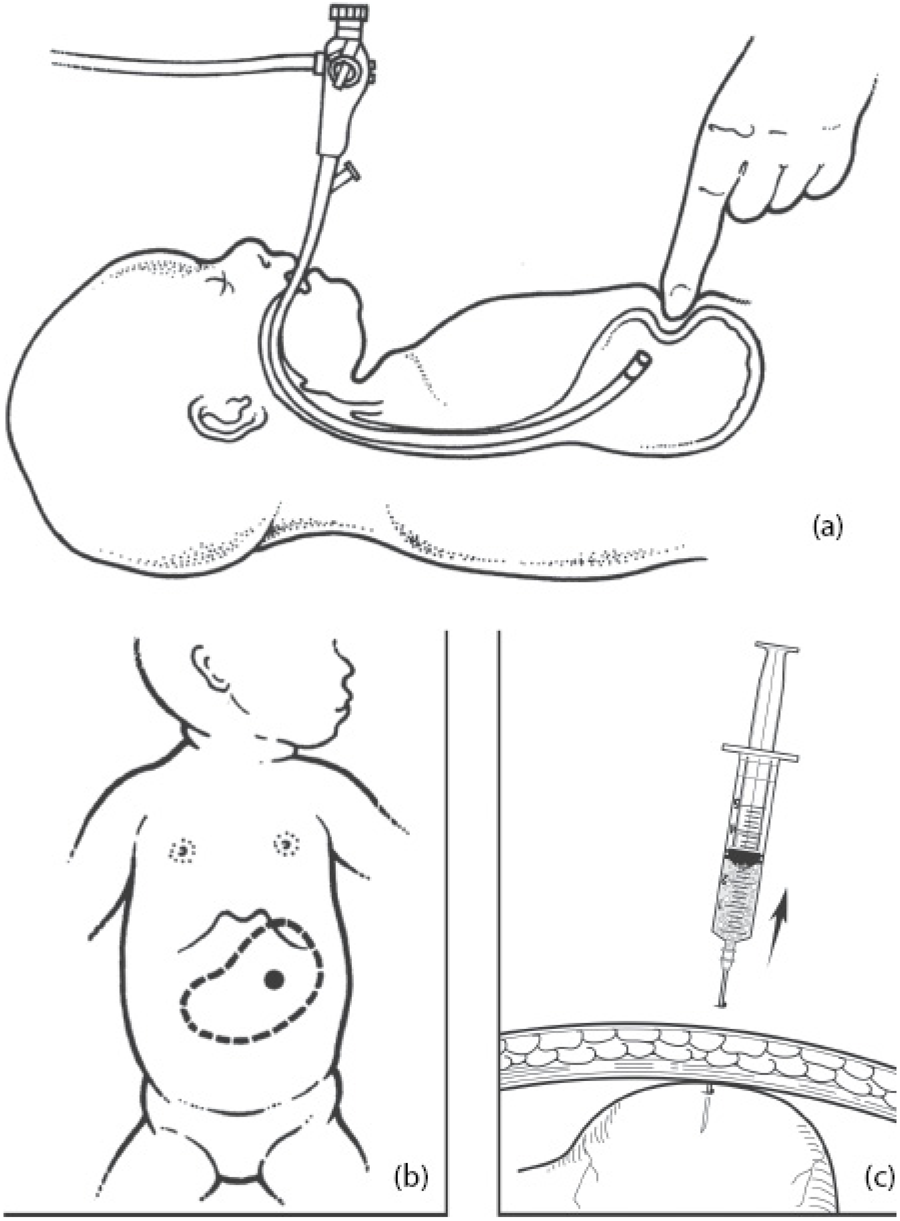
The operative field is prepared in the usual sterile manner. A pediatric catheter with a gastric retainer, markings on the shaft, and a dilating tapered end with a steel wire loop should be used (**Figure 31.9**).



31.9

The gastroscope is inserted and the stomach insufflated. The stoma should be away from the ribcage. Under- or overinsufflation should be avoided to minimize the possibility of accidentally piercing the transverse colon. Insufflation of the small intestine tends to push the transverse colon in front of the stomach and should be avoided.

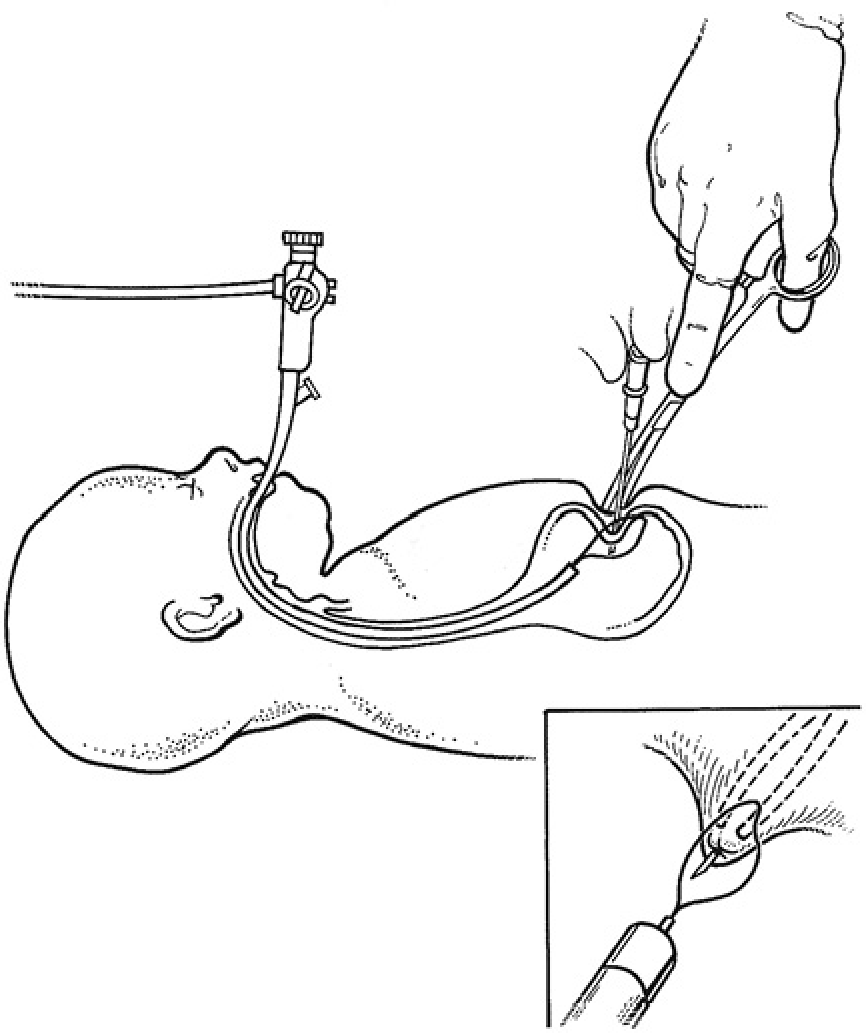
Digital pressure is applied to the proposed gastrostomy site, which usually corresponds to the area where transillumination is brightest (**Figure 31.10a-c**). Transillumination and clear visualization of an anterior gastric wall indentation are key moments in this procedure. Without these, an open or laparoscopic technique should be employed.



31.10a–c

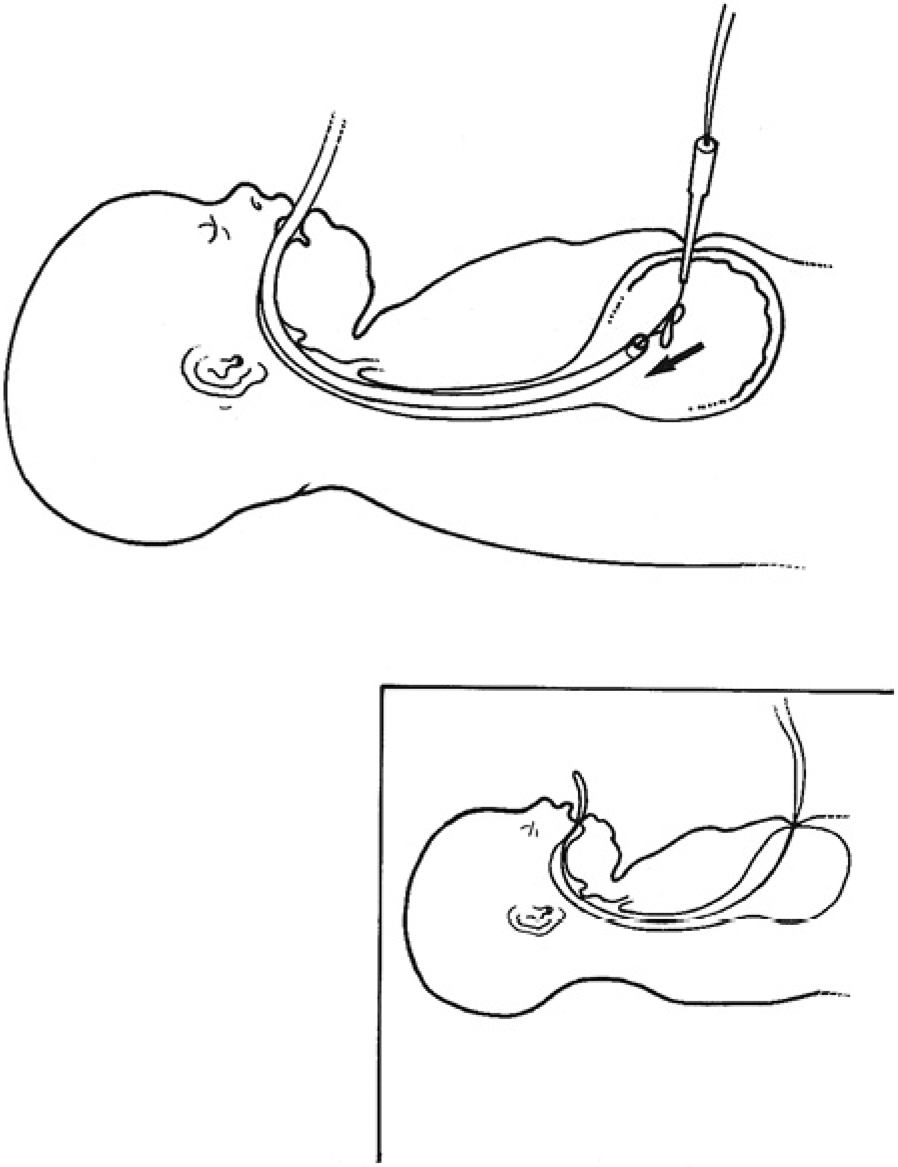
Local anesthetic is drawn into a syringe and the proposed PEG site is injected. The needle is advanced further and continuous aspirating pressure is applied to the plunger. Air bubbles should be visible in the remaining fluid when the tip of the needle is seen by the endoscopist. If air bubbles are noticed before the needle tip is in the stomach, the colon or other intestinal loop may be interposed between the stomach and the abdominal wall.

An 8–10 mm skin incision is made and a hemostat applied to maintain the intragastric indentation. The endoscope is moved in small increments. The endoscopist then places the polypectomy snare around this visible indentation. The IV cannula is placed in the incision between the prongs of the hemostat, and then firmly placed through the abdominal and gastric walls, exiting through the loop of the polypectomy snare (**Figure 31.11**). The snare is partially closed, but not tightened around the cannula.



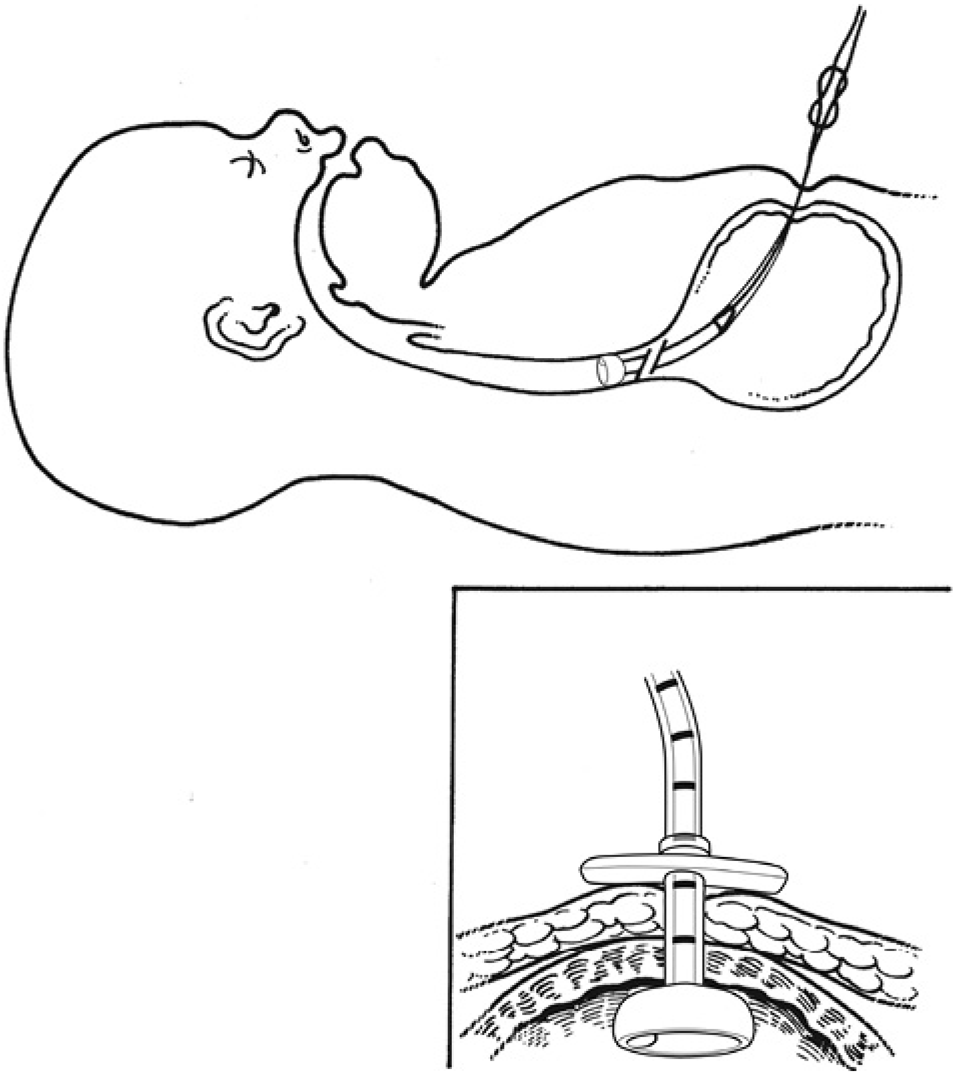
31.11

The needle is removed, and the looped steel wire is inserted through the cannula. The polypectomy snare is allowed to slide away from the cannula and is tightened around the wire. An alternative method is to retrieve the wire with alligator or biopsy forceps. The wire is then pulled back with the endoscope through the stomach and esophagus, exiting through the patient's mouth (**Figure 31.12**). A guiding tract is thus established.



31.12

The catheter is attached to the guidewire by interlocking the two steel wire loops. Traction is applied to the abdominal end of the wire, guiding the catheter through the esophagus and stomach and across the gastric and abdominal walls. The collapsed gastric retainer minimizes the risk of esophageal injury. (For diagrammatic purposes, a shortened catheter is shown in **Figure 31.13**.) The tapered end of the catheter exits through the abdominal wall before the gastric retainer enters the patient's mouth, allowing complete control of the catheter during placement. Traction is continued until the gastric and abdominal walls are in loose contact. The external crossbar or disk is slipped over the catheter and guided to the skin level, avoiding pressure from the retaining crossbar on the mucosa or skin. The catheter is cut to the desired length and the feeding adapter attached. No sutures are used, and the catheter is connected to a small, clear plastic trap. A gauze pad and tape are applied.

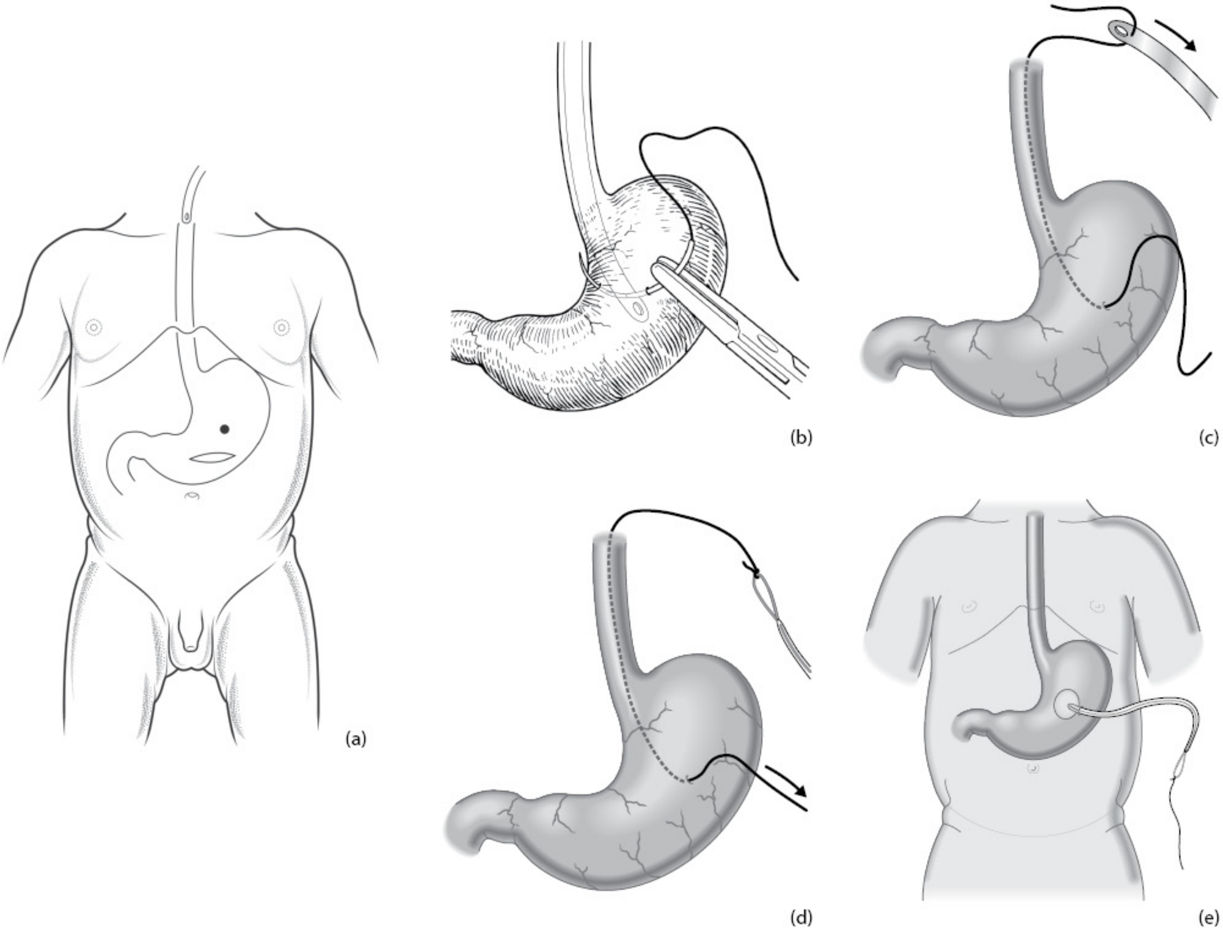


31.13

Hybrid gastrostomy

In children who have a markedly abnormal epigastric anatomy or dense adhesions because of previous abdominal operations or peritonitis, it may be impossible to perform a PEG or a laparoscopic gastrostomy safely. At times, even a Stamm gastrostomy may be technically challenging. For these cases, a hybrid procedure was developed that combines the "open" technique employing a mini-laparotomy with the "pull" PEG insertion of a gastrostomy catheter.

A mini-laparotomy is made in the left upper quadrant (**Figure 31.14a**). The anterior surface of the stomach is identified. A large (20–26 Fr) Nelaton-type catheter is inserted orally and advanced into the stomach, where it is clearly identified. A sturdy monofilament suture on a tapered needle is selected. The needle is pushed through the anterior gastric wall at the desired gastrostomy site, through the catheter, and then out of the stomach (**Figure 31.14b**). The needle is then cut off. The catheter is pulled back with the suture embedded in it (**Figure 31.14c**). The gastric end of the suture is brought out through a counter-incision (**Figure 31.14d**). The oral end of the suture is attached to a PEG-type catheter, which is then pulled back in a manner similar to the "pull" PEG illustrated above. **Figure 31.14e** demonstrates the completed procedure.



31.14a–e

Laparoscopic approaches to gastrostomy

Nowadays, most pediatric surgeons utilize minimally invasive techniques for placement of gastrostomy tubes. A significant advantage of laparoscopic over endoscopic techniques is that they allow for precise identification of the gastrostomy site on both the abdominal and the stomach walls.

Location on the abdominal wall should be chosen about two to three fingerbreadths away from the costal margin, as they tend to migrate cranially as the child grows, especially in neurologically devastated patients. The goal should be to place the stoma as low as possible without tension. Special caution should be taken to avoid injury to the epigastric vessels in the left upper quadrant.

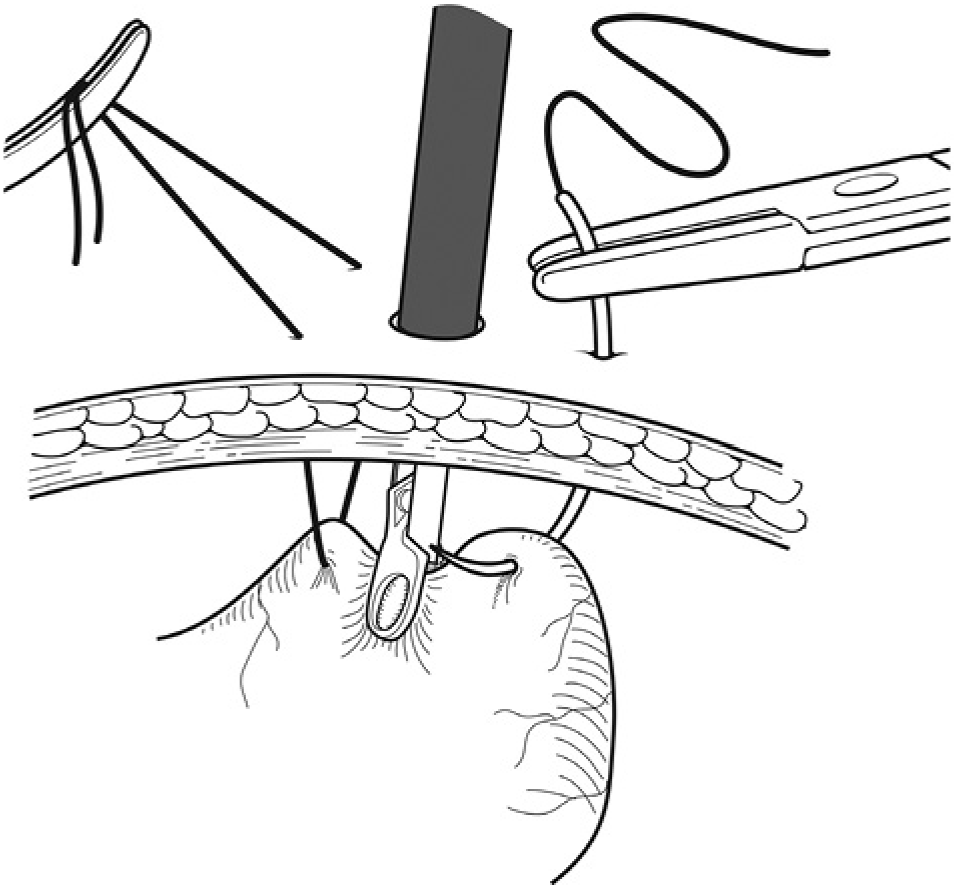
Location on the stomach wall should be at a dependent portion near the greater curvature, as stomas close to the pylorus can cause gastric outlet obstruction, and location closer to the lesser curvature complicates further Nissen fundoplication. Insertion at the greater curvature may theoretically worsen gastroesophageal reflux by opening the angle of His.

There are two broad types of laparoscopic approaches:

- *Laparoscopic percutaneous (Georgeson) gastrostomy:* Abdominal wall placement of stay sutures facilitates percutaneous access to gastrostomy site using serial dilations in a standard Seldinger technique.
- *Laparoscopic Stamm-type gastrostomy:* Placement of stay sutures fixed to fascia is followed by direct placement of the tube into the stomach through a seromuscular tunnel created by purse-string sutures.

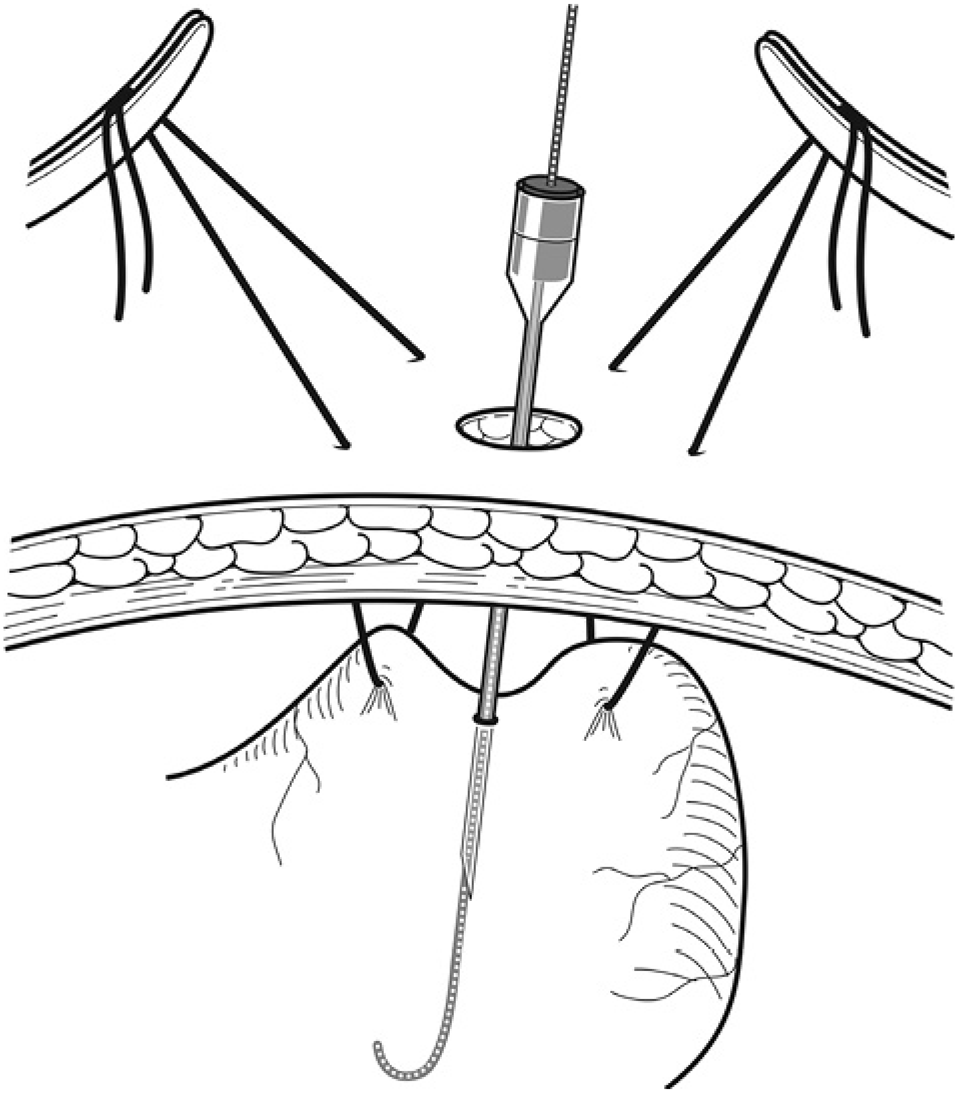
LAPAROSCOPIC PERCUTANEOUS (GEORGESON) GASTROSTOMY

The most suitable site of a gastrostomy is selected and marked. A nasogastric tube is inserted. Pneumoperitoneum is established in the usual, age-appropriate manner, and a 30-degree laparoscope is introduced at the umbilicus. The left epigastric area is inspected. The previously marked site is infiltrated with a long-lasting local anesthetic. The needle is then pushed through the abdominal wall and the appropriate relation between the anterior gastric wall and the stoma site established. A small skin incision is made, and a 5 mm trocar inserted. A bowel grasper is introduced and the stoma site on the anterior gastric wall is lifted toward the parietal peritoneum. In order to anchor the stomach to the abdominal wall temporarily, T-fasteners or U-stitches may be employed. The double, separate U-stitch technique is illustrated in **Figure 31.15**. A U-stitch is passed through the abdominal wall, through the anterior gastric wall, and back out through the abdominal wall. A second U-stitch is passed parallel to the first one, 1–2 cm apart. The sutures are lifted, maintaining the stomach in apposition with the abdominal wall.



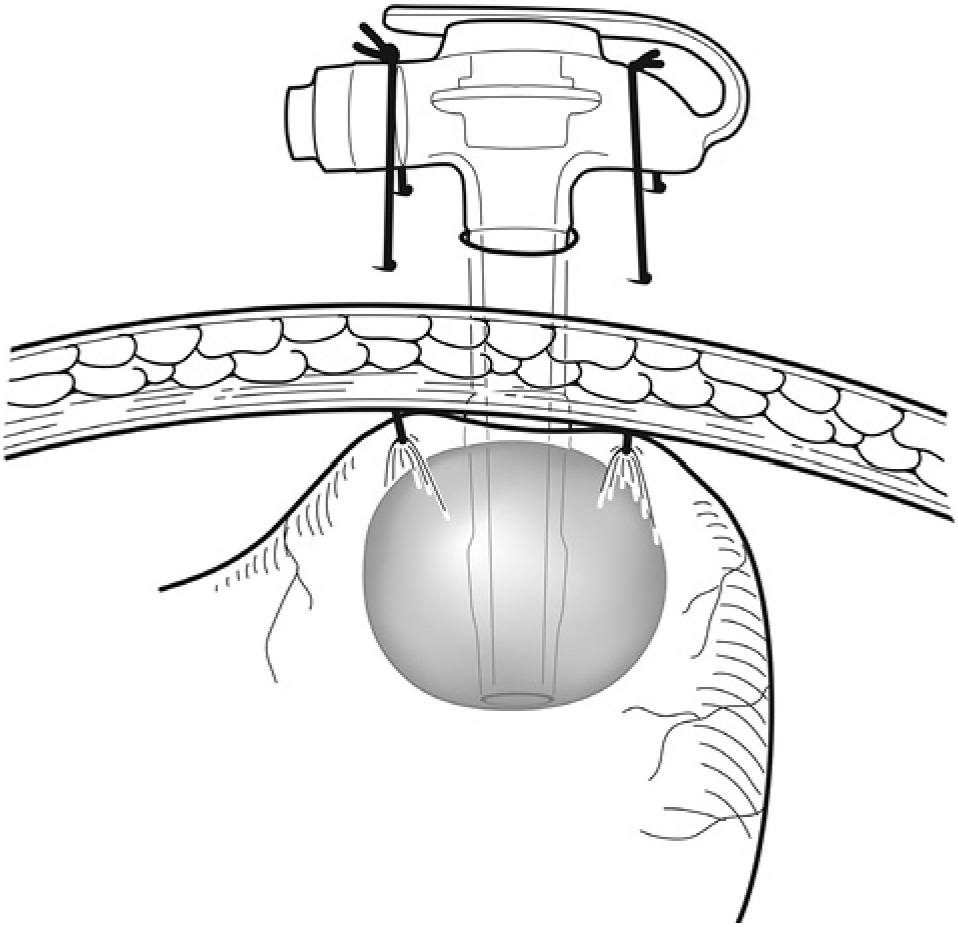
31.15

The stomach is insufflated with air through the nasogastric tube and a needle is inserted through the trocar site into the gastric lumen, between the two U-stitches. A guidewire is passed through the needle into the stomach. The tract is sequentially dilated over the guidewire with tapered dilators (8, 10, 16, and 20 Fr) to the size required to insert the gastrostomy device over the same guidewire (**Figure 31.16**). Stiffening of the catheter shaft with a thin dilator is helpful during this insertion.

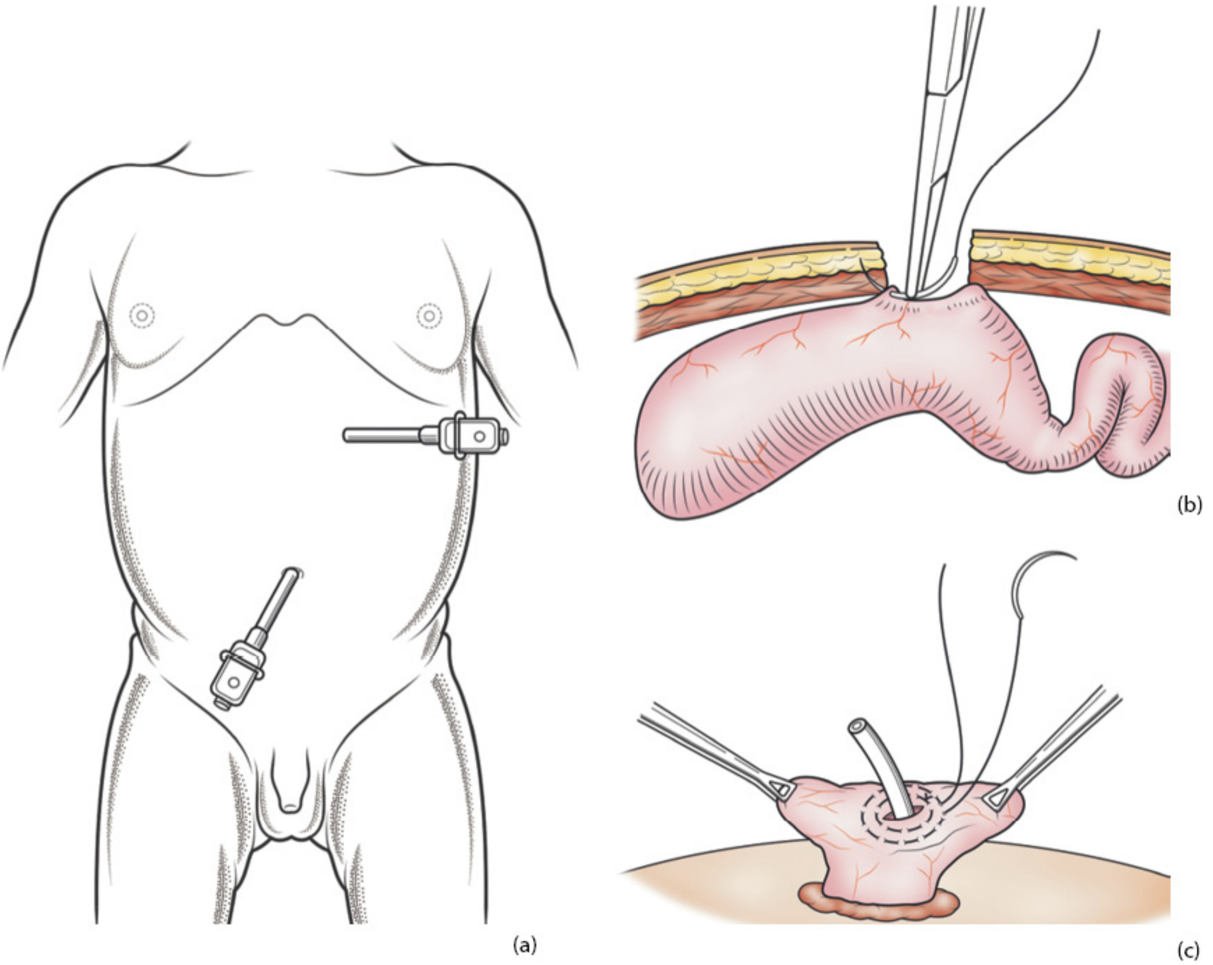


31.16

A balloon-type "button" is introduced and inflated under direct visualization to ensure adequate placement. The dilator and wire are removed. The previously placed U-stitches are tied over the wings of the "button" securing the gastric wall to the parietal peritoneum and the device to the abdominal wall (**Figure 31.17**). These sutures are to be removed 3–5 days postoperatively.



31.17



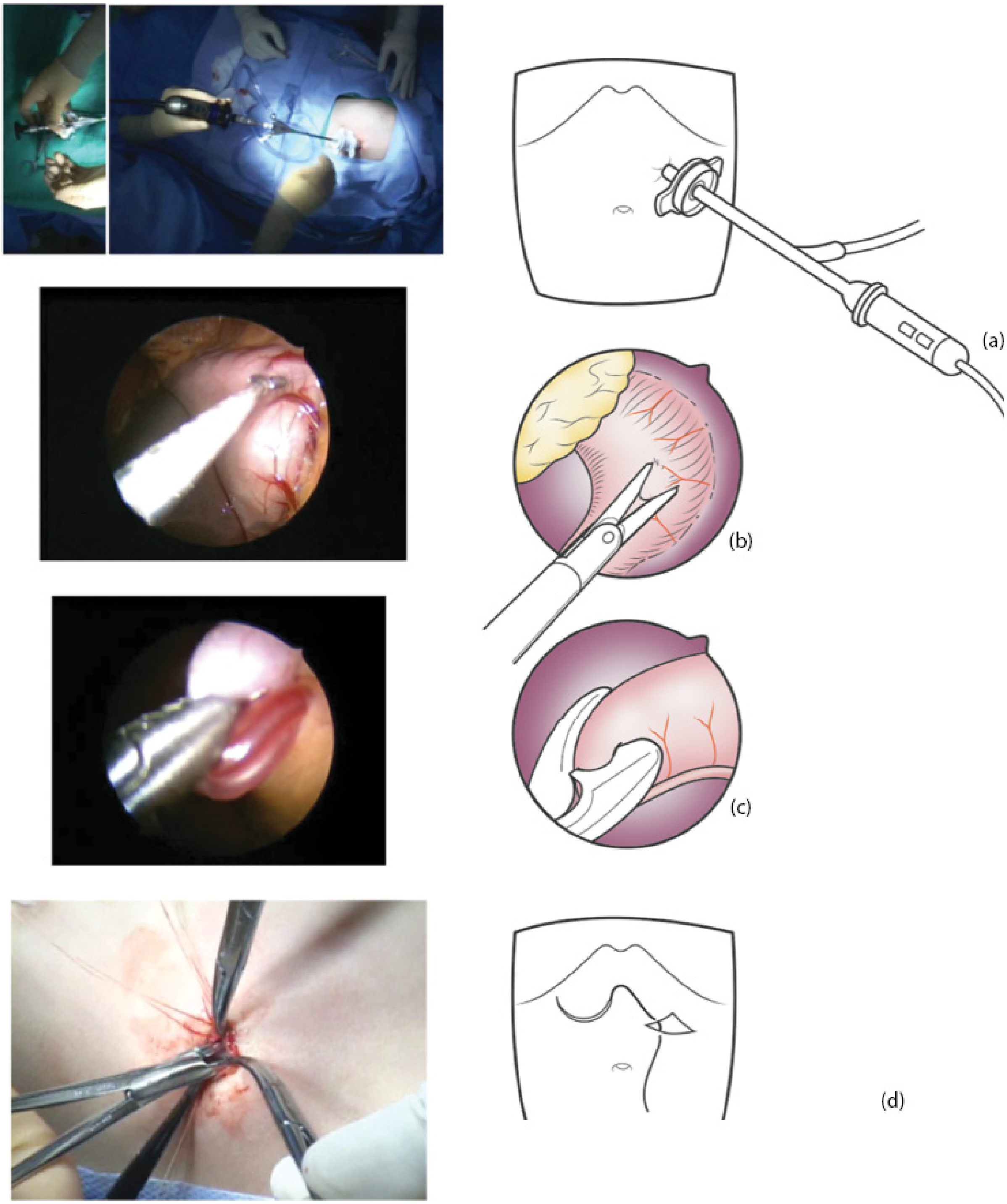
31.18a–c

LAPAROSCOPIC STAMM-TYPE GASTROSTOMY

A similar approach involves tacking the stomach directly to the fascia. This is typically accomplished with trocars at the umbilicus and the chosen site for the gastrostomy (**Figure 31.18a**). A laparoscopic Babcock□ or similar grasper is placed into the trocar and a site along the greater curvature, away from the pylorus, is grasped and brought through the abdominal wall. A stay suture is placed to allow manipulation of the site. The stomach is then secured directly to the fascia in three or four locations (**Figure 31.18b**), with a purse-string suture placed circumferentially leaving adequate room for the gastrostomy (**Figure 31.18c**). A gastrostomy is then made, and the gastrostomy tube is placed directly into the stomach. We typically inflate the balloon under direct laparoscopic visualization in order to confirm intraluminal placement. The purse string is then secured around the gastrostomy, and the tube is flushed to ensure that there is no leak.

LAPAROSCOPIC SINGLE-INCISION APPROACH

A single-incision approach has also been described. This technique is a simple variation of the above technique but obviates the need for an umbilical incision by using a 5 mm incision at the site of the intended gastrostomy in the left upper quadrant. A Veress needle is used to gain access, and a 5 mm trocar is placed after insufflation (**Figure 31.19a**). An optical grasper is then inserted through the trocar. The authors prefer to use a 5 mm rigid hysteroscope, which allows both visualization and manipulation through a single site. The stomach is visualized, with identification confirmed through insufflation via an indwelling orogastric tube. The stomach is grasped along the greater curvature (**Figure 31.19b**) and brought through the incision (**Figure 31.19c and d**). The gastrostomy is matured as described above, and intraluminal placement can be confirmed by infusing betadine–saline solution through the gastrostomy and aspirating it through the indwelling orogastric tube.



31.19a–d Laparoscopic single-incision approach.

One advantage of these fascial fixation techniques is that they allow primary placement of a gastrojejunostomy tube (GJ-tube), which can be a temporary or permanent alternative to an antireflux procedure.

There are three ways to place a post-pyloric jejunostomy tube in this situation:

- The tube can be advanced manually into the jejunum with direct visualization through an orally placed endoscope (often using guidewires).
- The tip of the tube can be guided endoscopically by pulling the string at the tip of the jejunal catheter past the pylorus.
- Using fluoroscopy, guidewires can be passed through the gastrostomy site to make the c-loop around the duodenum, with the GJ-tube then placed over the wire.

Percutaneous gastrostomy and gastrojejunostomy guided by fluoroscopy, computed tomography, or ultrasonography

These techniques are, in many ways, similar to the previously described methods of creating an approximation of the anterior gastric wall to the parietal peritoneum and inserting a self-retaining tube. For the fluoroscopic and computed tomography (CT)-guided procedures, the stomach is insufflated with air; in the ultrasonographic-guided gastrostomy, the stomach is filled with saline. The stomach may be approximated and held in place by T-fasteners prior to the insertion of the catheter or a pigtail catheter may be used to gain access and retention. In either case, a Seldinger-type technique is employed, followed by dilatation of the tract and catheter insertion.

The primary placement of GJ-tubes is becoming more common. With just minor modifications from a standard gastrostomy, they can help temporarily avoid fundoplication. Available as an image-guided percutaneous procedure performed by interventional radiologists, GJ-tubes can be inserted in patients with factors that are prohibitive for endoscopic access or general anesthesia. After the gastric lumen is identified with fluoroscopy, CT scan, or ultrasound, gastropexy is accomplished with T-fasteners. Gastric puncture is accomplished with the needle aimed toward the pylorus to allow passage of a rigid catheter into the duodenum through which a guidewire is passed beyond the ligament of Treitz under fluoroscopic guidance. The skin tract is dilated in a similar way to other percutaneous techniques, and a GJ-tube catheter is inserted over the guidewire with position in the proximal jejunum confirmed with contrast administration.

POSTOPERATIVE CARE

Enteral feedings begin following open gastrostomies once the ileus has resolved, and on the day after the operation for minimally invasive procedures. Some groups have described “fast-track” or same day feeds for gastrostomy tubes.

Our dressing preference is to leave the gastrostomy uncovered. We avoid harsh antiseptic solutions and, after a few days, simply use soap and water for cleaning.

Granulation tissue tends to form after a couple of weeks and is controlled with applications of silver nitrate or triamcinolone cream. If granulation tissue becomes excessive, it leads to leakage and needs to be excised. We have observed good results with the application of triamcinolone and antifungal combination to prevent the recurrence of granulation tissue. Once the tract becomes epithelialized, no medication should be used.

If a “button” has not been placed at the initial procedure, the long tube should be replaced with one of the skin-level devices as soon as a reliable adherence of the stomach to the abdominal wall has occurred, typically around 4–6 weeks postoperatively.

Should a gastrostomy no longer be needed, the catheter is simply removed. The tract will usually close in a matter of days. If the gastrocutaneous fistula remains after several weeks or is causing skin excoriation due to leakage of gastric contents, closure of the tract is performed.

COMPLICATIONS

Although generally considered a basic procedure, gastrostomy is associated with a long list of complications related to technique, care, and catheter use. Serious technique-related problems include separation of the stomach from the abdominal wall leading to peritonitis, wound separation, hemorrhage, infection, injury to the posterior gastric wall or other organs, and placement of the tube in an inappropriate position. Separation of the stomach from the abdominal wall is usually due to inadvertent, premature dislodgement of the tube or a disruption during catheter change. It requires immediate attention. Interventional approaches can be used to salvage, although laparoscopic correction may be possible in select cases. Most complications can be avoided by careful choice of the procedure and stoma device and using meticulous technique, approximating the stomach to the abdominal wall, and avoiding tubes in the midline or too close to the ribcage.

Among the most common long-term problems are leakage and granulation tissue. Severe leakage is initially managed using conservative measures and interrogation of the GJ-tube to ensure the balloon is appropriately inflated and not leaking. If these fail, or if there is severe prolapse of gastric mucosa, the stoma may need to be revised or relocated.

FOLLOW-UP

All children with gastrostomies should be carefully followed up to prevent long-term catheter-related complications and monitored for manifestations of foregut dysmotility, particularly gastroesophageal reflux.

VIDEO

OPS_8e_video_31.1 Gastroscopy via single incision

FURTHER READING

Aprahamian CJ, Morgan TL, Harmon CM, et al. U-stitch laparoscopic gastrostomy technique has a low rate of complications and allows primary button placement: Experience with 461 pediatric procedures. *J Laparoendosc Adv Surg Tech A* 2006; 16: 643–9.

Gauderer MWL. Experience with a hybrid, minimally invasive gastrostomy for children with abnormal epigastric anatomy. *J Pediatr Surg* 2008; 43: 2178–81.

Gauderer MWL, Ponsky JL, Izant RJ Jr. Gastrostomy without laparotomy: A percutaneous endoscopic technique. *J Pediatr Surg* 1980; 15(6): 872–5.

Gauderer MWL, Stellato TA. Gastrostomies: Evolution, techniques, indications and complications. *Curr Probl Surg* 1986; 23: 661–719.

Ponsky JL, Gauderer MWL. Percutaneous endoscopic gastrostomy: A nonoperative technique for feeding gastrostomy. *Gastrointest Endosc* 1981; 27(1): 9–11.

Ponsky TA, Lukish JR. Single site laparoscopic gastrostomy with a 4-mm bronchoscopic optical grasper. *J Pediatr Surg* 2008; 43(2): 412–14.

Rothenberg SS, Bealer JF, Chang JH. Primary laparoscopic placement of gastrostomy buttons for feeding tubes. A safer and simpler technique. *Surg Endosc* 1999; 13(10): 995–7.

32

Pyloromyotomy

NIGEL J. HALL

HISTORY

Sabricius Hildanus first described pyloric stenosis in 1646. Harald Hirschsprung elaborated on the clinical presentation and pathology of the condition in 1888. At this stage, the preferred treatment was medical, using a combination of gastric lavage, antispasmodic drugs, dietary manipulation, and the application of local heat, because the surgical mortality was almost 100%. In 1908, Fredet advocated longitudinal submucosal division of the thickened pyloric muscle, but recommended suturing the defect transversely. In 1912, Ramstedt simplified the Fredet procedure by omitting the transverse suturing, leaving the mucosa exposed in the longitudinal subserosal defect. This operation was successful and its essential elements have remained virtually unmodified ever since. Surgery has now completely replaced medical measures for the treatment of pyloric stenosis.

The operative approach to the pyloromyotomy has also undergone a number of modifications in the last few decades. Up until the 1990s, the majority of surgeons approached the pylorus through either a transverse or a midline incision in the upper abdomen. Frequently, the cosmetic result was unsatisfactory. In 1986, Tan and Bianchi reported the use of a paraumbilical incision to approach the pylorus with superior cosmesis and this has become the standard open approach in many centers. More recently, laparoscopic pyloromyotomy has been reported and has superseded an open procedure in many specialist pediatric surgical units.

PRINCIPLES AND JUSTIFICATION

Incidence

The incidence of pyloric stenosis among white people is 2–3 per 1000 live births. Black infants are less frequently affected. The male-to-female ratio is 4 : 1. The disorder often occurs in firstborn boys, and there is a strong familial pattern of inheritance. Vomiting due to pyloric stenosis has been noted in twins whose symptoms began within hours of each other.

Diagnosis

Symptoms usually commence at 2–4 weeks of age, but can sometimes be seen in neonates or infants close to 2 months of age. Symptoms consist of projectile vomiting of non-bilious material, constipation, dehydration, lethargy, or seizures, and failure to thrive. Hematemesis has been documented in a few cases.

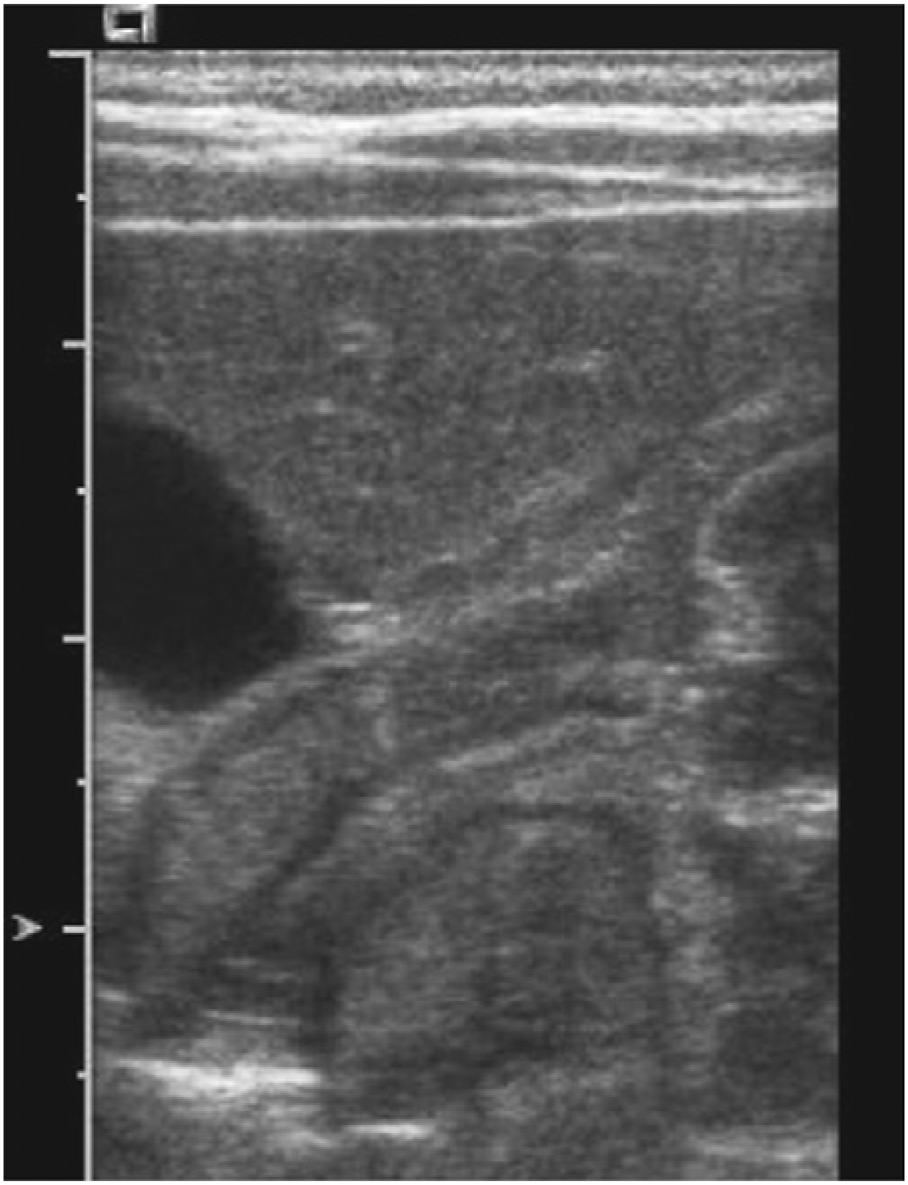
Physical signs include variable degrees of dehydration, visible gastric peristalsis, and a palpable pyloric tumor.

If one takes the time to perform a proper clinical examination, the diagnosis can nearly always be made without further investigations, but more often patients will be referred to the surgeons with diagnostic images suggestive of pyloric stenosis.

While diagnostic images may be helpful in difficult cases, none is necessary if clinical examination demonstrates a palpable “tumor” or “olive.” When physical examination fails to identify a pyloric mass, ultrasonographic images should be obtained. If ultrasound is equivocal, barium swallow can be helpful but is rarely required. Endoscopy should not be necessary to make the diagnosis of pyloric stenosis, but it may be useful in symptomatic patients with other causes for gastric outlet obstruction.

ULTRASONOGRAPHY

Ultrasonography typically shows a thickened pyloric musculature with a central sonolucent area representing the lumen. The thickened muscle is easy to recognize as a donut or bull’s eye (**Figure 32.1**). Ultrasonographers also use the following measurement criteria to make the diagnosis: Pyloric channel >17 mm in length and pyloric thickness >4 mm.



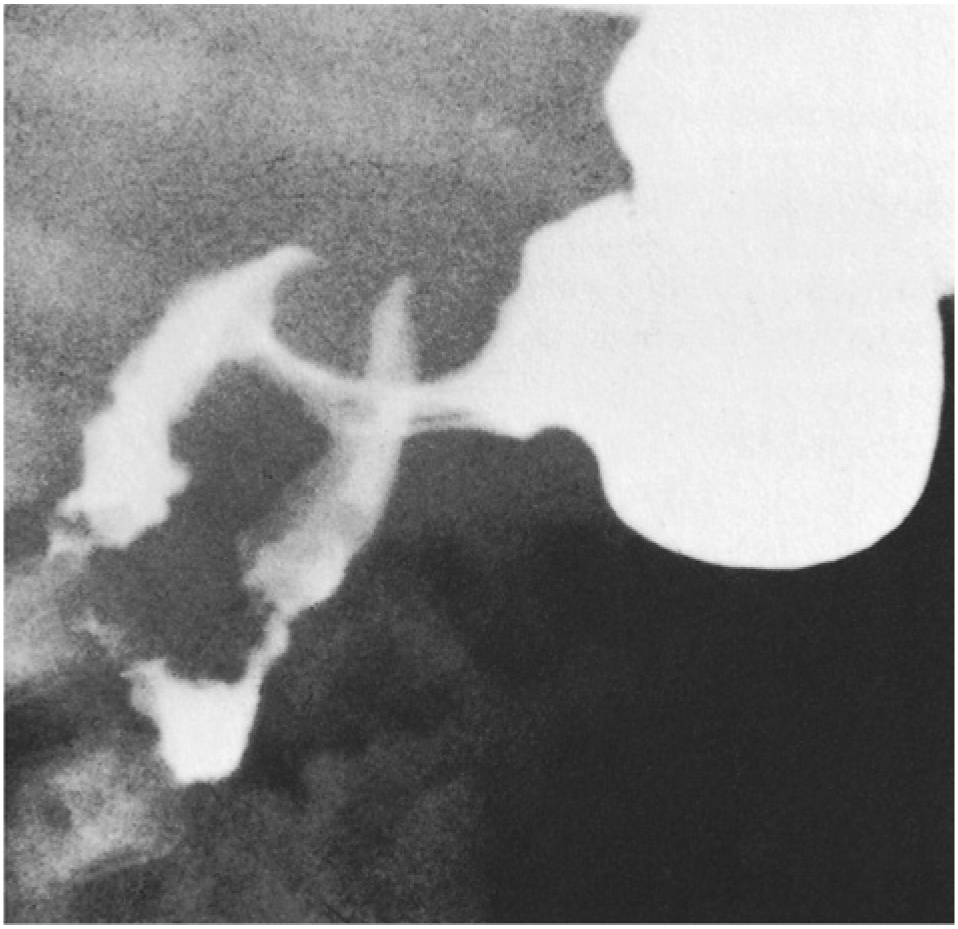
32.1

BARIUM SWALLOW

The following features are diagnostic (**Figure 32.2**):

- “String sign” of the narrow elongated pyloric canal
- “Double track” in the pyloric canal owing to infolding mucosa

- Delayed gastric emptying
- Gastric hyperperistalsis
- The mushroom effect in the duodenal cap due to indentation by pyloric "tumor"



32.2

PREOPERATIVE PREPARATION

Pyloromyotomy is never an emergency; correction of dehydration and acid–base imbalance takes precedence. Parenteral correction of metabolic abnormalities is the safest way to prepare a patient for surgery. An IV infusion of isotonic saline (20 mL/kg) is administered over 60 minutes. Once urinary output has been established, potassium chloride (20–30 mmol/L) is added to the infusate of 5% glucose in 0.45% saline. The infusion is given at a rate of 120–150 mL/kg body weight per 24 hours.

It may take as long as 48–72 hours for rehydration to be complete and for the infant to be ready for surgery. The goal is to correct the serum electrolytes to nearly normal. Accordingly, the serum potassium should be at least 3–4.5 mEq/L, the serum CO_2 should be <27–30 mEq/L, and the serum sodium should be >130 mEq/L before surgery is considered. The serum chloride will usually correct itself to >100 mEq/L with the above measures.

Preoperatively, a nasogastric tube should be inserted, allowed to drain freely, and aspirated every 4 hours. The volume of fluid drained should be replaced with an IV infusion of an equal volume of 0.9% saline containing 20 mmol/L potassium chloride.

OPERATION

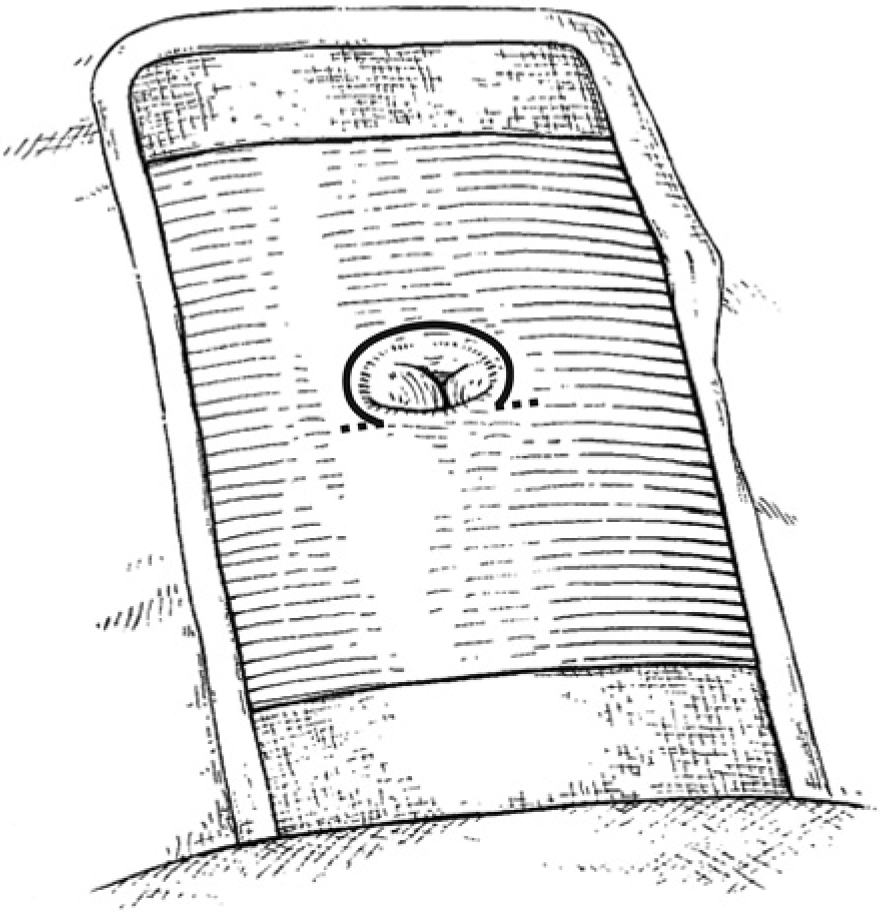
Open pyloromyotomy

POSITION OF PATIENT

The patient is placed supine on the operating table, prepared, and draped in the usual sterile fashion. A preoperative prophylactic antibiotic should be given at the time of the operation to minimize the risk of postoperative wound infection.

INCISION

A curvilinear incision approximately 2 cm long is made in the upper fold of the umbilicus, circumscribing the umbilicus itself (**Figure 32.3**). If the umbilicus is small, the incision may be extended laterally into an "omega" shape to improve access. The incision is deepened through the subcutaneous tissues down to the linea alba, which is stripped of overlying tissue longitudinally for approximately 2 cm above the umbilicus. The linea alba is then opened in a cephalad direction, the umbilical vein retracted to one side, and the peritoneum opened longitudinally.



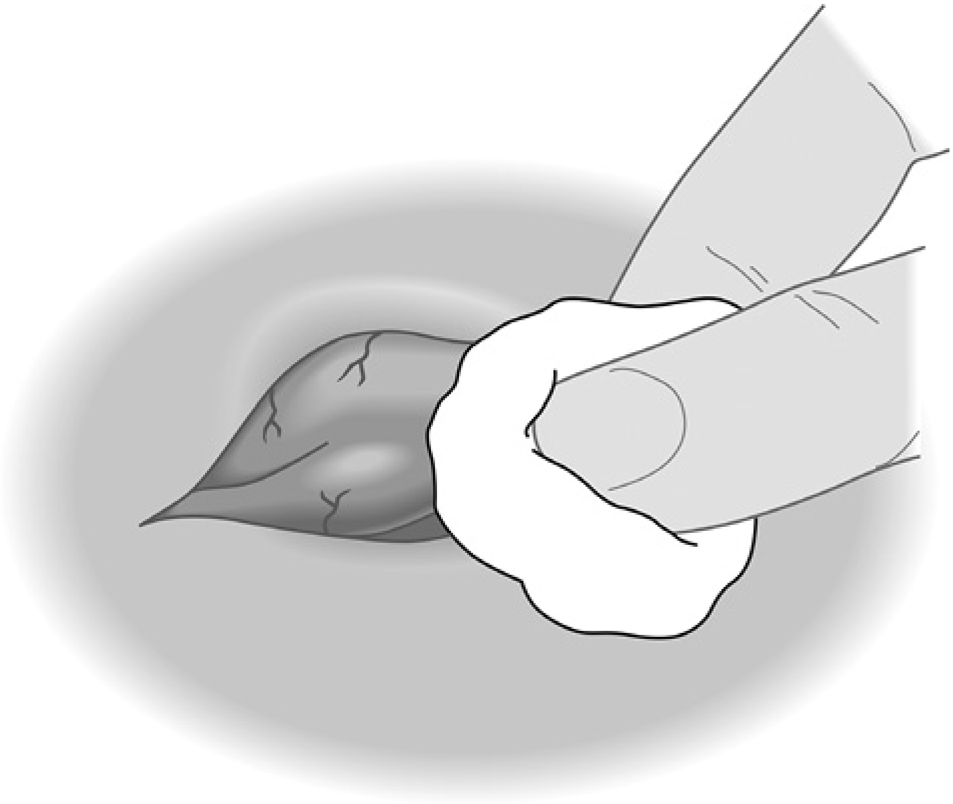
32.3

IDENTIFICATION OF THE STOMACH

With the peritoneum opened, the stomach may be visible but is often not. If it is, it is grasped with non-traumatic forceps and delivered gently into the wound where it is held with a moist gauze swab. No attempt should be made to grasp the pyloric tumor directly, as this leads to serosal tears and hemorrhage. If the stomach is not visible, gentle traction on the transverse colon or greater omentum will allow identification of the greater curve of the stomach.

DELIVERY OF THE PYLORIC TUMOR

With the greater curvature of the stomach firmly drawn across to the left and exerting traction on the antrum, the pyloric tumor is delivered out of the incision by applying a gentle to-and-fro rocking traction on the pylorus (**Figure 32.4**). If this proves difficult, the likely cause is inadequate length of the skin incision. Extension of the incision will reduce the risk of serosal tearing and aid delivery of the pyloric tumor.



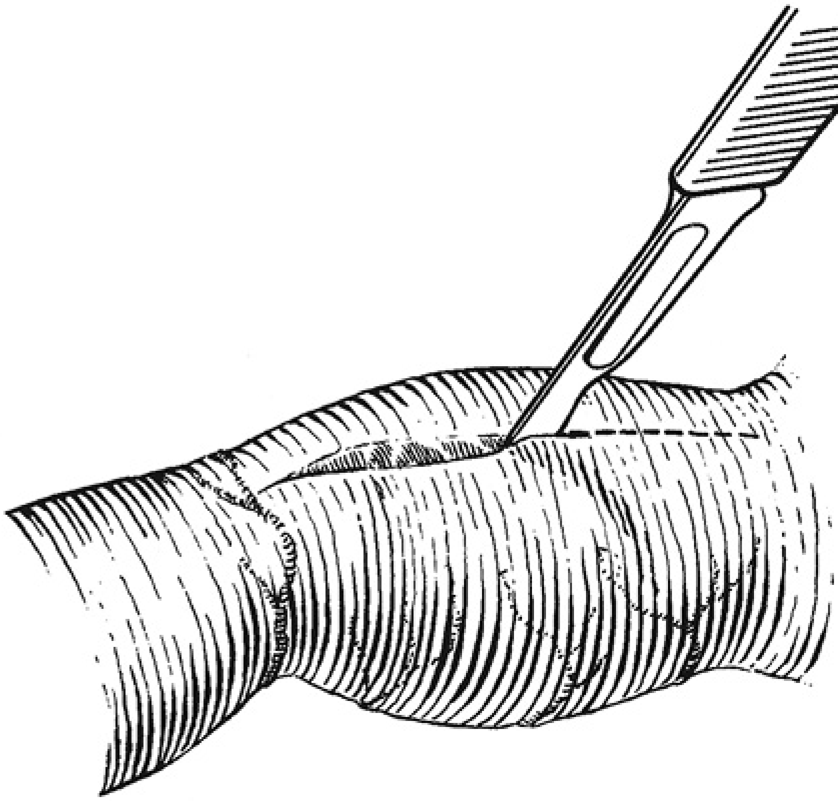
32.4

The pyloric vein of Mayo marks the distal extent of the tumor. Proximally, the tumor is less obvious where it merges with the hypertrophied stomach musculature. The tumor has a glistening, grayish appearance and is firm to palpation.

There is a relatively avascular plane in the middle of the anterior surface where the vessels entering the pylorus superiorly and inferiorly merge.

INCISION OF THE PYLORUS

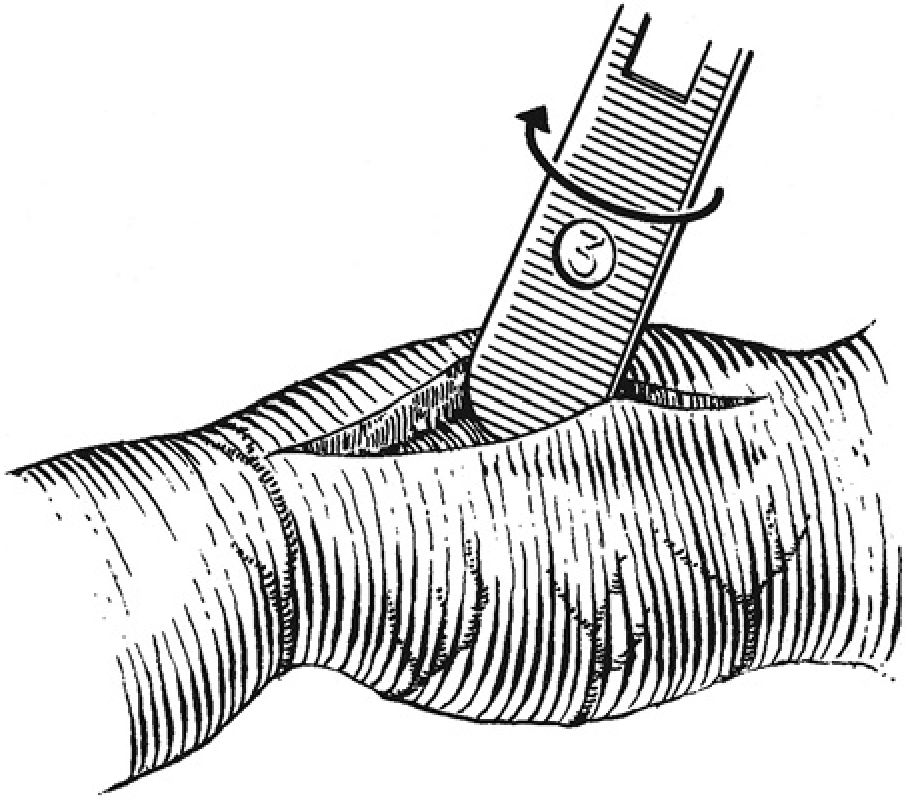
A serosal incision is then made in the avascular area on the anterior surface of the tumor (**Figure 32.5**). It is carried distally as far as the pyloric vein of Mayo, which marks the pyloroduodenal junction, while proximally it extends well onto the anterior surface of the antrum of the stomach. The length of the incision is 2–3 cm.



32.5

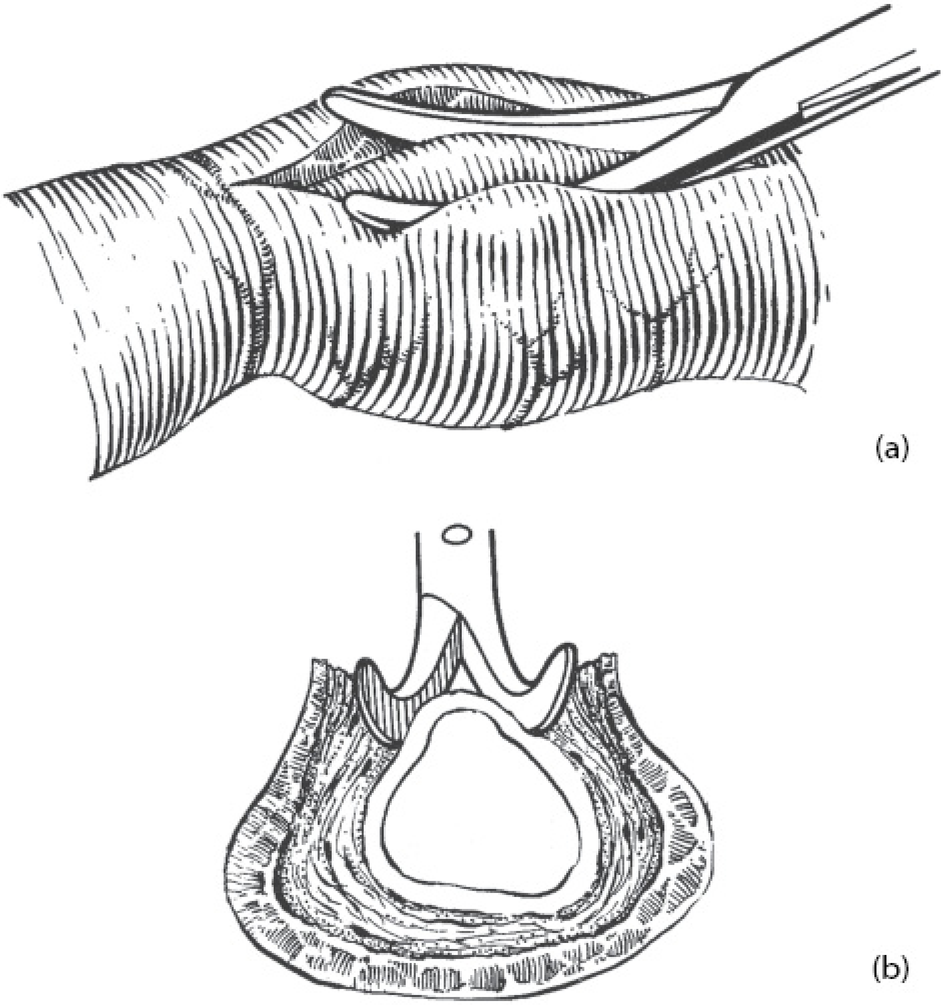
SPLITTING OF THE PYLORIC MUSCULATURE

Pressure with a blunt instrument, such as the back of the handle of a scalpel, into the incision with counterpressure from a finger placed behind the tumor allows splitting of the hypertrophied muscle fibers down to the submucosa (**Figure 32.6**). This appears as a white, glistening membrane in the depth of the incision of the pylorus. A twisting movement on the blunt instrument produces an extension of the split proximally and distally and widens the incision. Alternatively, the blunt instrument is gently rubbed back and forth along the incision and over the muscle to split it.



32.6

To ensure that all muscle fibers have been divided throughout the length of the incision, the edges of the split muscle are spread apart with either a pair of blunt forceps (ensuring that the points are held away from the mucosa) or a pyloric spreader (Dennis Browne or Benson and Lloyd) so that the submucosa bulges into the incision (**Figure 32.7a and b**). Special care must be taken at the pyloroduodenal junction to avoid entering the lumen of the duodenum, which is particularly vulnerable because of the protrusion of the pyloric tumor into the duodenal lumen. The adequacy of the split is assured if the two halves of the muscle move independently of each other.



32.7a,b

TESTING FOR PERFORATION

About 10–20 mL of air is introduced into the stomach via the nasogastric tube and then gently milked through the pylorus into the duodenum, and a gauze sponge is dabbed on the incision to detect any bile staining. Any perforation of the mucosa will become obvious at this juncture and should be closed by direct suture with 6/0 polydiaxonone or polyglycolic acid. Some surgeons advocate closing the pyloromyotomy completely and redoing the myotomy on the opposite side of the pylorus. In either case, the important point is to recognize the perforation and to repair any leak found.

Slight hemorrhage from the edges of the pyloromyotomy will cease once the tumor has been replaced into the peritoneal cavity and hence venous congestion relieved.

WOUND CLOSURE

The wound is closed with an interrupted suture of polyglycolic acid or polydiaxonone for peritoneum/muscle and fascia. The subcutaneous tissues are closed with continuous 4/0 polyglycolic acid suture and the skin is approximated with 5/0 polyglycolic acid subcuticular suture or surgical glue.

Laparoscopic pyloromyotomy

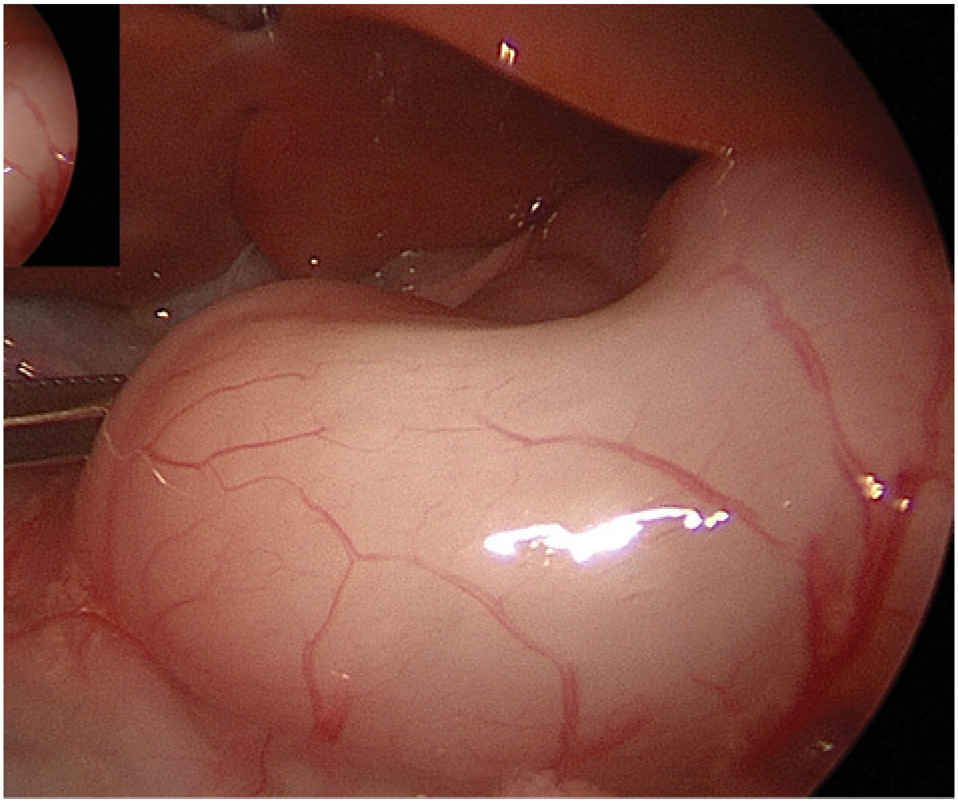
HISTORY

In 1991, Alain et al. described laparoscopic extramucosal pyloromyotomy for the first time. Since then, several other groups have reported their experience with the technique for laparoscopic extramucosal pyloromyotomy. However, the application of laparoscopy to the treatment of hypertrophic pyloric stenosis occurred late compared to other procedures in the pediatric population, as it was difficult to justify using it rather than the open technique. Tan and Najmaldin reported their initial experience in 1993 and 1995, with good results. Several high-quality prospective randomized controlled trials have now been performed comparing open and laparoscopic approaches.

OPERATIVE PROCEDURE

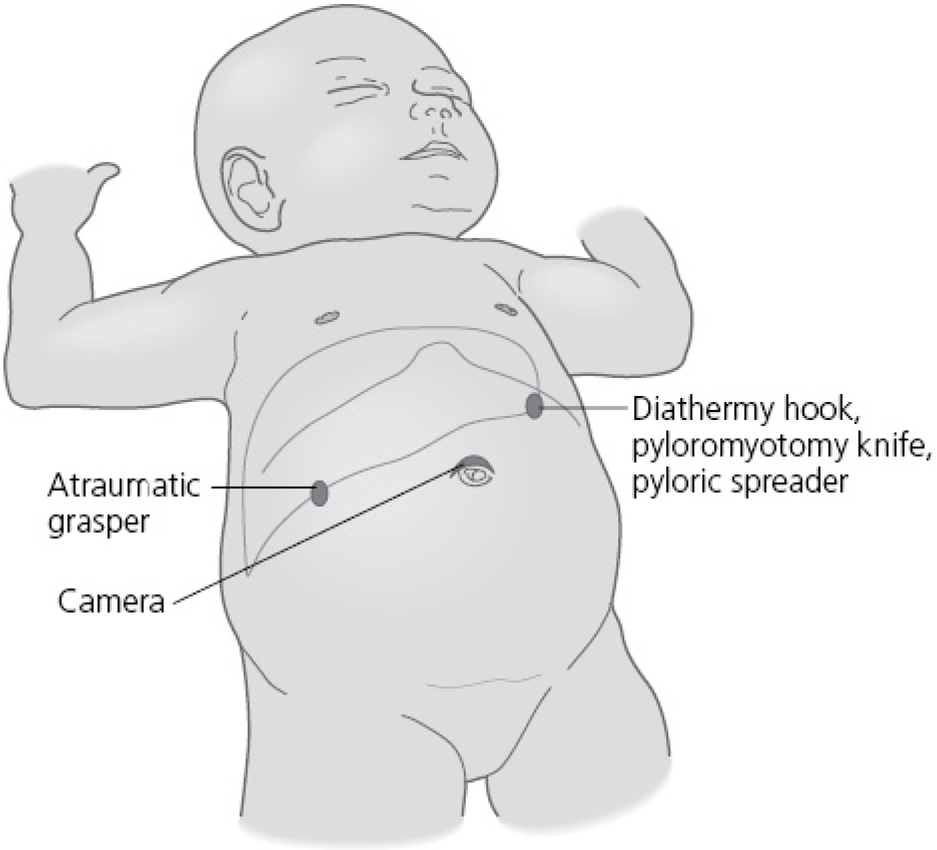
After the induction of anesthesia, the patient is placed in the supine position with the feet at the very bottom of the operating table and the table shortened if possible. The laparoscopic screen should be positioned over the patient's head facing the feet. The abdomen is prepared and draped in the usual sterile fashion, with special attention being paid to meticulous preparation of the umbilicus. A preoperative prophylactic antibiotic should be given at the time of the operation to minimize the risk of postoperative wound infection.

The surgeon and assistant should stand on either side of the patient initially. A stab incision is made in the supraumbilical skin crease using a no. 11 knife blade, and stretched along the contour lines to allow the passage of a 5 mm trocar. The subcutaneous tissues are divided to expose the linea alba which is opened followed by the peritoneum under direct vision. A purse-string-type absorbable suture is inserted around the defect, but not tied. A 5 mm trocar is inserted, the purse-string suture tightened to provide an airtight seal, and a single throw of a knot placed on the suture to secure the port in position. A pneumoperitoneum is established keeping the pressure as low as possible (ideally 6–8 mmHg). A small-caliber 30-degree telescope is placed through the umbilical port. A general abdominal inspection is performed and the thickened pylorus is visualized (**Figure 32.8**).



32.8

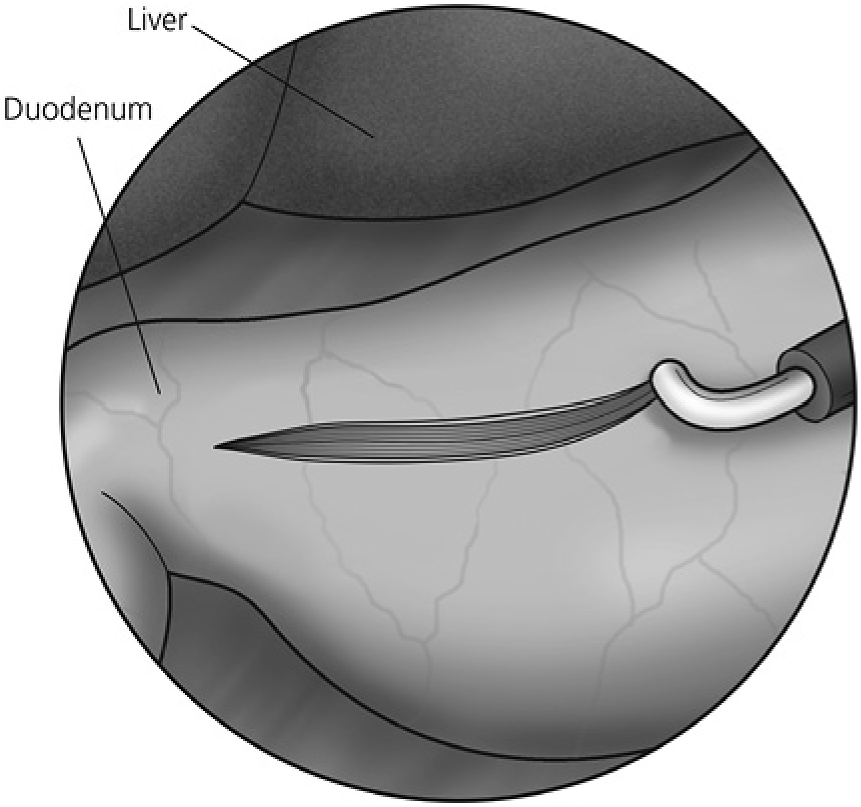
A small stab incision is made on the left side of the abdomen lateral to the mid-clavicular line, halfway between the umbilicus and the costal margin, and a small-caliber trocar is introduced for an atraumatic grasper passed directly through the abdominal wall (**Figure 32.9**). This is used to stabilize the pylorus either by supporting the duodenum distally or by grasping the stomach proximally. Another stab incision is made on the right side of the abdomen in a similar position but slightly higher on the abdomen. A 3 mm diathermy hook is passed into the abdomen to initiate the pyloromyotomy. Some surgeons prefer to use a retractable arthroscopic knife instead.



32.9

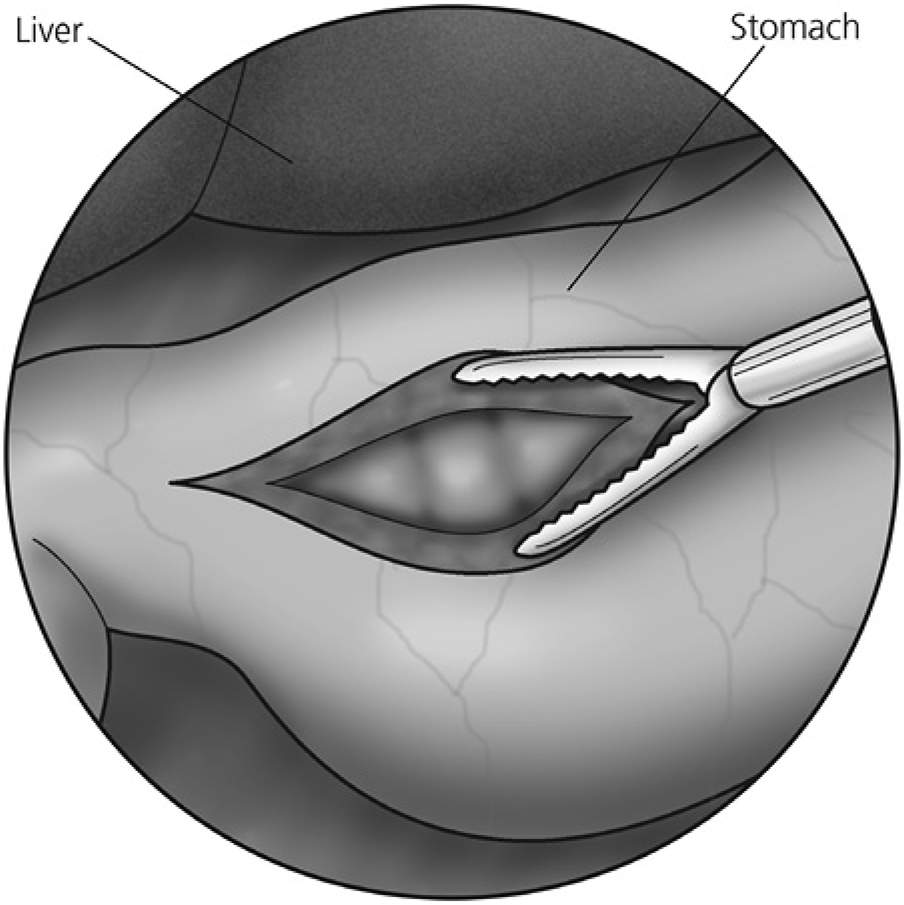
The pyloromyotomy is made using monopolar diathermy or the knife in an area of avascularity over the pylorus (**Figure 32.10**). The incision should extend from the anterior gastric wall to just proximal to the vein of Mayo.

The knife or diathermy hook is then withdrawn and a laparoscopic spreader introduced. The pyloric incision is deepened bluntly under vision as in the open technique to expose the bulging mucosa.



32.10

The pyloromyotomy in **Figure 32.11** shows the split hypertrophied pyloric muscle with exposed intact mucosal layer in the depth of the split.



32.11

An adequate myotomy is assessed by holding the edges of the myotomy using an atraumatic grasper and pyloric spreader and moving them independently to be certain that the two halves of the pylorus move independently.

The stomach should then be insufflated with air through a nasogastric/orogastric tube and milked through the pylorus into the duodenum to be certain that there are no leaks and that the air passes easily.

Following completion of the procedure, the stab incisions are closed with surgical glue. The supraumbilical port is removed, and the purse-string suture loosened and then retied to close the wound. The skin is closed with glue.

Postoperative analgesia can be achieved by infiltrating the closed wounds with 0.25% bupivicaine.

POSTOPERATIVE CARE

Patients are transferred from the recovery room to a regular hospital bed and IV fluids are continued. Feedings are begun and advanced as per surgeon preference. The authors find that, whereas in the past surgeons fed infants slowly, most of the time these infants can be advanced in their feedings quite rapidly once it is clear that they are tolerating oral intake, and start with an initial feeding of 15 mL of the infant's regular milk. If that is tolerated we commence *ad libitum* feeds 1 hour later. It is not unusual to see some occasional postoperative emesis. If this persists, it is usually due to curds of milk retained in the stomach, in which case, the authors lavage the stomach with a 10% solution of sodium bicarbonate and begin the feeding regimen again.

COMPLICATIONS

Other than the occasional postoperative emesis, the most frequently observed complication is wound infection. This is usually due to *Staphylococcus aureus* and is less frequently seen with the laparoscopic approach.

Mucosal perforation may occur as previously described. Following closure of the perforation, a nasogastric tube is left following the procedure and IV antibiotics given for 48 hours. The infant remains "nil by mouth" for 24 hours postoperatively. If they remain well after this time, feeds are commenced. Incomplete pyloromyotomy is a rare complication (approximately 1%) and is due to an inadequate length of pyloromyotomy, usually at the gastric end. After initial improvement, the vomiting persists. This complication requires distinction from gastroesophageal reflux and the diagnosis is made by upper gastrointestinal contrast study. Repeat pyloromyotomy (open or laparoscopic) is curative.

VIDEOS

OPS_8e_video_32.1 Laparoscopic pyloromyotomy

FURTHER READING

Adibe OO, Nichol PF, Lim FY, Mattei P. Ad libitum feeds after laparoscopic pyloromyotomy: A retrospective comparison with a standardized feeding regimen in 227 infants. *J Laparoendosc Adv Surg Tech* 2007; 17: 235–7.

Alain JL, Grousseau D, Terrier G. Extramucosal pyloromyotomy by laparoscopy. *Surg Endosc* 1991; 5: 174–5.

Hall NJ, Pacilli M, Eaton S, et al. Recovery after open versus laparoscopic pyloromyotomy for pyloric stenosis: A double-blind multicentre randomised controlled trial. *Lancet* 2009; 373: 390–8.

Leclair MD, Plattner V, Mirallie E, et al. Laparoscopic pyloromyotomy for hypertrophic pyloric stenosis: A prospective, randomized controlled trial. *J Pediatr Surg* 2007; 42: 692–8.

Najmaldin A, Tan HL. Early experience with laparoscopic pyloromyotomy for infantile hypertrophic pyloric stenosis. *J Pediatr Surg* 1995; 30: 37–8.

St Peter SD, Holcomb GW III, Calkins CM, et al. Open versus laparoscopic pyloromyotomy for pyloric stenosis: A prospective, randomized trial. *Ann Surg* 2006; 244: 363–70.

Tan KC, Bianchi A. Circumumbilical incision for pyloromyotomy. *Br J Surg* 1986; 73: 399.

33

Bariatric surgery

LINDEL C.K. DEWBERRY AND THOMAS H. INGE

HISTORY

Pediatric obesity is increasingly recognized as a significant healthcare problem. With the lack of effective medical therapy, the need for surgical intervention has been established. As the epidemic of pediatric obesity has been increasing and the efficacy and safety of bariatric surgery for adults has also become evident, more consideration has been given to bariatric procedures for severely obese adolescents. Over the past 30 years, many bariatric procedures with various modifications have been introduced, including intestinal bypass, loop gastric bypass, horizontal and vertical gastroplasty, Roux-en-Y gastric bypass (RYGBP), biliopancreatic diversion, adjustable gastric banding, and, most recently, sleeve gastrectomy. From the beginning of the subspecialty of bariatric surgery, the goal of the operation has been to restrict the intake of nutrients, interfere with the absorption of nutrients that are ingested, or both. The long-term goal is to maintain a degree of weight reduction which improves or eliminates obesity-related comorbidities and decreases the risk of future obesity-related medical complications and death.

PRINCIPLES AND JUSTIFICATION

Currently, approximately 9% of adolescents meet the definition of severe obesity and will likely remain severely obese as adults. Significant, durable weight loss and long-term medical benefits have now been documented for adults who have undergone bariatric surgery. For this reason, current opinion holds that bariatric surgery should also be an option for highly selected adolescents with extreme obesity. Since relatively little is known about the long-term sequelae of bariatric surgery when performed in adolescence, conservative indications for operation have been recommended. Surgery should be reserved after at least 6 months of organized weight-loss attempts have failed.

Recent best practice guidelines have been developed for pediatric/adolescent weight loss surgery. These new guidelines more closely mimic those for adult surgery than in previous years. Those considered for weight loss surgery include adolescents with a body mass index (BMI) ≥35 kg/m^2 and several specific obesity-related comorbidities for which there is clear evidence of short-term morbidity (type 2 diabetes, severe steatohepatitis, pseudotumor cerebri, and moderate to severe obstructive sleep apnea). Those adolescents with a BMI ≥40 kg/m^2 and other long-term medical comorbidities should also be considered for weight-loss surgery. Physical maturity should be documented by either history and physical examination or radiographic study, thus generally limiting surgery to those over the age of 12 years. The reader is referred to other sources for further discussion of the contraindications and preoperative decision making for adolescent bariatric surgery.

Bariatric surgery effectively creates a “tool” with which adolescents can lose one-third or more of their body mass and concomitantly reduce or eliminate most comorbidities of adolescent obesity. Currently, there are two minimally invasive techniques being used for all of the modern weight loss procedures in adolescents. These include RYGBP and the sleeve gastrectomy (SG). The procedure that has been used most commonly in this country and worldwide is the RYGBP, as this procedure has been associated with the safest side-effect profile, balanced with excellent long-term efficacy and maintenance of weight loss.

Bariatric programs for adolescents should include expertise in adolescent obesity, nutrition, diet, and behavioral management. It is critical that thorough investigations be conducted to discover unrecognized coexisting obesity-related medical conditions. Minimally invasive surgery has significant advantages over open surgery, including a reduction in both length of hospitalization and operative morbidity. Minimally invasive bariatric surgery is one of the most technically difficult operations to perform. Laparoscopic skills utilized in foregut surgery are not directly transferable to bariatric surgery. Expertise in minimally invasive surgery may not confer the same level of expertise in performing minimally invasive bariatric surgery. Surgeons performing bariatric procedures must be well trained, as suggested by the American Society for Bariatric Surgery, the American College of Surgeons, and the Society of American Gastrointestinal Endoscopic Surgeons. Prior to performing laparoscopic bariatric operations, surgeons must meet all local credentialing requirements for the performance of open bariatric procedures and advanced laparoscopic operations. Given the controversy centered on adolescent bariatric surgery, the aspiring adolescent bariatric surgeon, at a minimum, should take a course in bariatric surgery and perform his or her first five to ten procedures proctored by an experienced laparoscopic bariatric surgeon.

PREOPERATIVE ASSESSMENT AND PREPARATION

The preoperative education of the patient, family, and, to some extent, the referring pediatrician is of paramount importance. Information about the surgical procedure, alternative operations, and the rationale for the operation to be performed, postoperative care, and the considerable lifestyle modifications required afterwards must be conveyed verbally and in writing to the patient and caregivers. The adolescent and parents must demonstrate understanding of the details of the procedure, its known and potential risks, and predictable consequences, and that understanding should be reflected in the archived responses to a written test. The referring pediatrician will need to become familiar with the anatomic and physiologic changes that occur after operation and develop an understanding of common postoperative complications to effectively partner with the surgeon in the care of the patient for years following the operation.

In addition to standard preoperative labs, micronutrient deficiency screening is performed in order to provide preoperative supplementation of vitamin D, iron, and other vitamins if needed. On the day prior to surgery, the patient is limited to clear liquids. No bowel preparation is required. Preoperative medications include low molecular weight heparin (40 mg s.c. and continued twice a day postoperatively while in hospital), and a second-generation cephalosporin (2 g IV). Our prefererence is cefoxitin. Sequential compression boots are also used intra- and postoperatively and should be applied and functioning prior to induction of anesthesia. All patients should be considered candidates for the laparoscopic approach unless they have had extensive abdominal surgery in the past.

Anesthesia

Patients undergo general anesthesia for all laparoscopic bariatric surgical procedures. There is generally no need for epidural analgesia. Routine preoperative consultation with the anesthesiologist and cardiologist is recommended to uncover potentially important information about the patient's airway and the patient's or family's anesthetic history, and to determine whether occult cardiovascular disease is present. Preoperative anesthesia consultation also provides an opportunity for unhurried discussion of any concerns or questions the family or patient may have about the technique and risks of anesthesia.

Equipment

Although extra-long (45 cm) laparoscopic instrumentation is available for bariatric procedures, laparoscopic bariatric procedures can generally be performed using standard 32 cm adult instrumentation. Various manufacturers supply the equipment required and the choice of one over another is largely a matter of surgeon preference. Specific instrumentation used at our institution for these procedures includes the following:

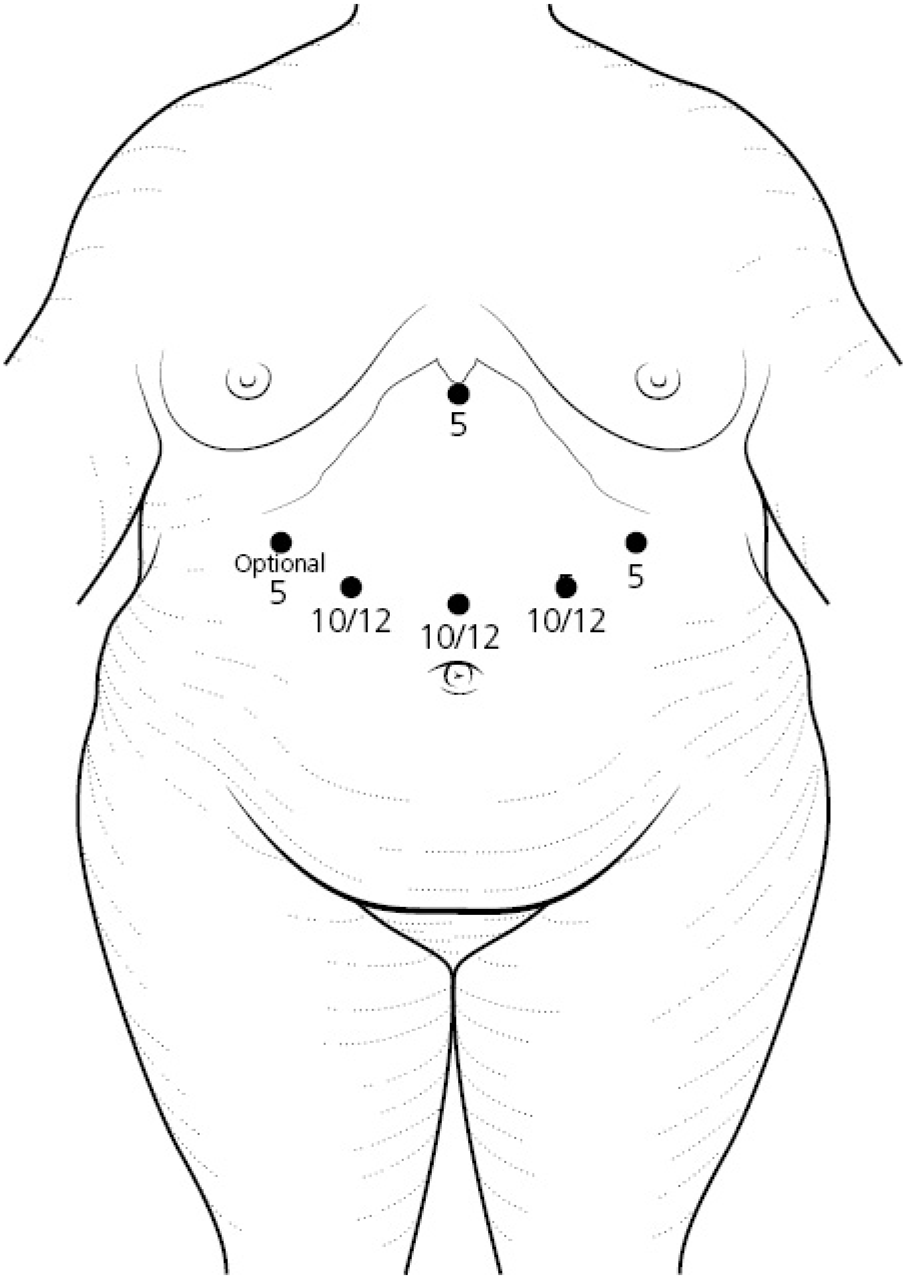
- All procedures:

  - 1% lidocaine, 10 mL syringe, and a long (spinal) needle
  - zero-degree 32 cm laparoscope (for initial abdominal access) with ENDOPATH□ XCEL□ Optiview trocar (Ethicon Endosurgery, Cincinnati, OH, USA)
  - 30-degree 32 cm laparoscope
  - Locking atraumatic bowel grasper
  - 2 × non-locking atraumatic bowel graspers
  - Locking toothed grasper (for liver retraction)
  - Hook scissor (for cutting suture)
  - Needle driver
  - Wolfe bipolar electrocautery forceps
  - Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA)
  - Multiple 20 cm lengths of 2/0 silk and 3/0 dyed Vicryl□ on SH needles
- Specific to RYGBP:
  - 3 × 10 mm or 12 mm ENDOPATH□ XCEL□ trocars (Ethicon Endosurgery)
  - 2 or 3 × 5 mm ENDOPATH□ XCEL□ trocars (Ethicon Endosurgery)
  - 15 mm ENDOPATH□ XCEL□ trocar (Ethicon Endosurgery)
  - Echelon Flex□ 60 mm Endocutter stapler (Ethicon Endosurgery; part no. LONG60A)
  - Multiple blue (3.5 mm Ethicon part no. ECR60B) and white (2.5 mm Ethicon Endosurgery; part no. ECR60W) staple loads
  - Endoscopic curved circular stapler 25 mm (end-to-end anastomosis, EEA) (Ethicon Endosurgery, part no. ECS33)
  - Gore SEAMGUARD□ bioabsorbable staple line reinforcements, 25 mm EEA (WL Gore and Associates, Newark, DE, USA)
  - Anvil grasper
  - Articulating grasper (Richard Wolf, Vernon Hills, IL, USA)
  - Carter-Thomason CloseSure System XL□ (Cooper Surgical Inc., Trumbull, CT, USA; part no. CTXL)
  - Jackson Pratt flat drain
- Specific to SG:

- 2 × 10 mm or 12 mm ENDOPATH□ XCEL□ trocars (Ethicon Endosurgery)
- 3 × 5 mm ENDOPATH□ XCEL□ trocars (Ethicon Endosurgery)
- 15 mm ENDOPATH□ XCEL□ trocar (Ethicon Endosurgery)
- Echelon Flex□ 60 mm Endocutter stapler (Ethicon Endosurgery; part no. LONG60A)
- Green (4.1 mm Ethicon Endosurgery, part no. ECR60G) and multiple blue (3.5 mm Ethicon Endosurgery; part no. ECR60B) staple loads
- Multiple Gore SEAMGUARD□ bioabsorbable staple line reinforcements, 60 mm linear (WL Gore and Associates, Newark, DE, USA)
- Piece of umbilical tape measured to 6 cm in length
- 34 Fr orogastric tube (Kimberly Clark, Irving, TX, USA; part no. 15034)
- Carter-Thomason CloseSure System XL□ (Cooper Surgical Inc.; part no. CTXL)
- Jackson Pratt flat drain

LAPAROSCOPIC ROUX-EN-Y GASTRIC BYPASS OPERATIVE PROCEDURE

This procedure is primarily a "one surgeon" procedure, with the assistant's role largely that of tissue retraction. Thus, the experienced laparoscopic bariatric surgeon can perform this operation without another trained bariatric surgeon as an assistant. During laparoscopic RYGBP, the patient can be flat or in gentle reverse Trendelenburg position with legs together. The surgeon stands on the right, the scrub nurse to the surgeon's right side, and the assistant on the left side of the patient. Given the patient's extreme girth, it is ergonomically advantageous to stand on a platform 20–40 cm above the floor. For initial abdominal access, the transparent, bladeless, direct viewing (XCEL Optiview□, Ethicon Endosurgery, Cincinnati, OH, USA) 12 mm trocar can be used safely and efficiently. This trocar is placed through the midline approximately 5–10 cm above the umbilicus. The exact site for this trocar is determined by laying a standard 10 mm adult laparoscopic telescope on the patient's abdomen, with the tip of the scope (the end which will be inside) at the nipple level, marking a skin site at the point beside the light cord insertion point on the scope. This skin site will provide optimal visualization of the upper and lower portions of the procedure. The two other 12 mm and one or two 5 mm trocar placement sites and the 5 mm subxiphoid site for liver retraction are shown in **Figure 33.1**.

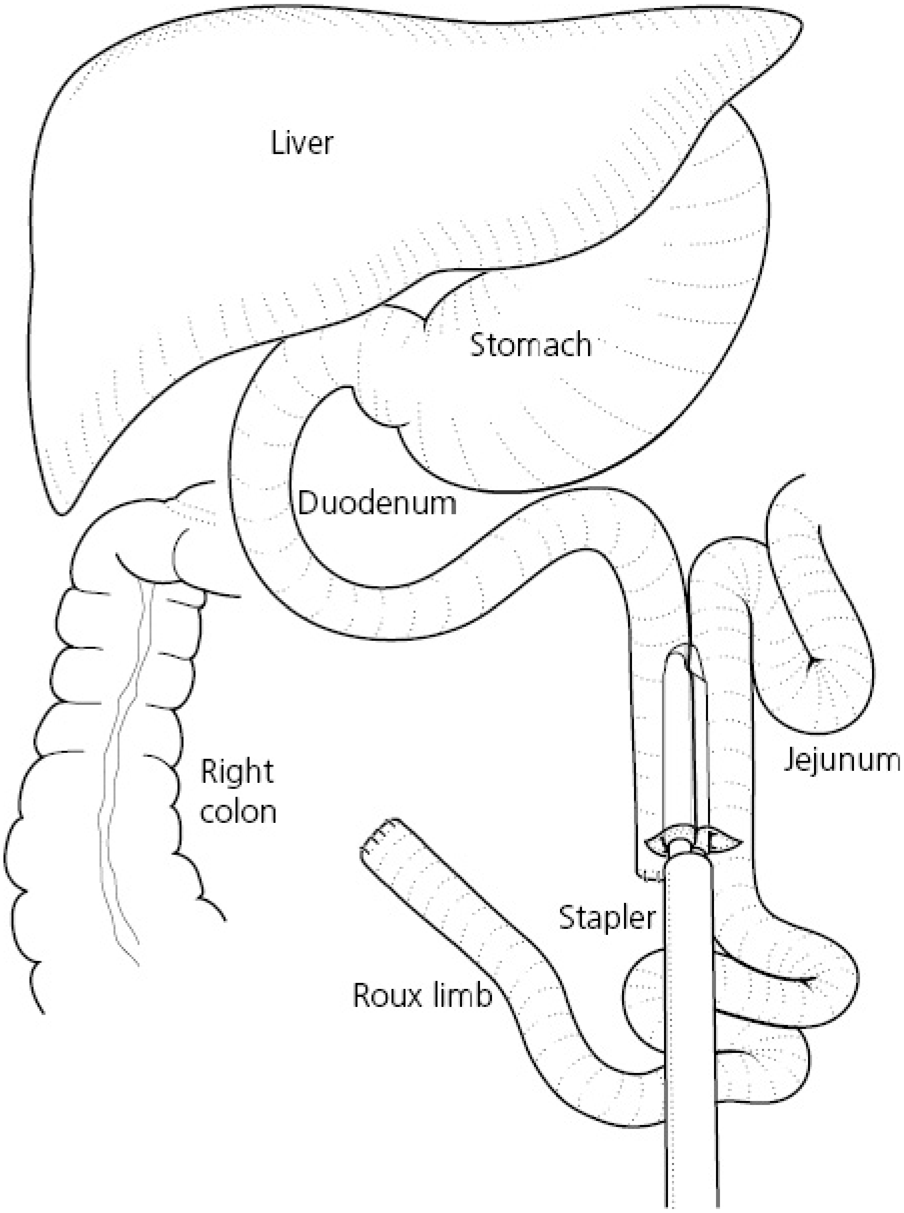


33.1

Roux limb construction

The omentum is first raised cephalad to expose the transverse mesocolon and origin of the jejunum (ligament of Treitz). Exposure of the small bowel is optimized if the table is placed into slight Trendelenburg position and the omentum is tucked carefully under the liver edge. The transverse mesocolon is grasped with a locking grasper just anterior to the duodenojejunal flexure and elevated anteriorly. The jejunum is divided generally 50 cm beyond the duodenojejunal flexure using the Echelon 60 mm linear stapler (EES, white load). The Roux limb is generally brought up to the pouch in the antecolic fashion. This requires at least 50 cm to reach the pouch in the epigastrium comfortably. The mesentery is minimally divided at this point using either half the length of a second stapler load or the Harmonic scalpel® (Ethicon Inc., Cincinnati, OH, USA). Clamping the stapler jaws together for a count of 10 seconds before firing mechanically thins the highly vascular tissue to reduce bleeding from the staple line. Bipolar electrocautery may be needed to achieve full hemostasis in this area.

There is no unanimity about the exact length of the Roux limb. Current practice employs a 150 cm Roux limb for most patients. With experience, the limb length can be visually estimated, by “walking” hand over hand down the bowel with laparoscopic graspers. Precision in measurement can be increased by using a 6 mm Penrose drain of known length alongside the bowel if desired. Next, a single stay suture is placed to approximate the antimesenteric borders of the distal end of the biliopancreatic segment (the proximal jejunum) and the point 150 cm distal to the end of the Roux limb. The Harmonic scalpel® (Ethicon Inc., Cincinnati, OH, USA) is used to make opposing enterotomies, which are spread wide enough with a grasper to accept the stapler jaw (Figure 33.2). A side-to-side jejunojejunostomy is created using the 60 mm Echelon stapler with white load. The resulting enterotomy defect is closed either with a running 3/0 Vicryl® (SH needle) or by using another firing of the stapler (preferred method). There is an increased risk of narrowing the lumen if the stapled closure is not applied precisely. The mesenteric defect is next closed with a running 2/0 silk suture to avoid an internal hernia orifice.


Liver
Stomach
Duodenum
Jejunum
Right colon
Stapler
Roux limb

33.2

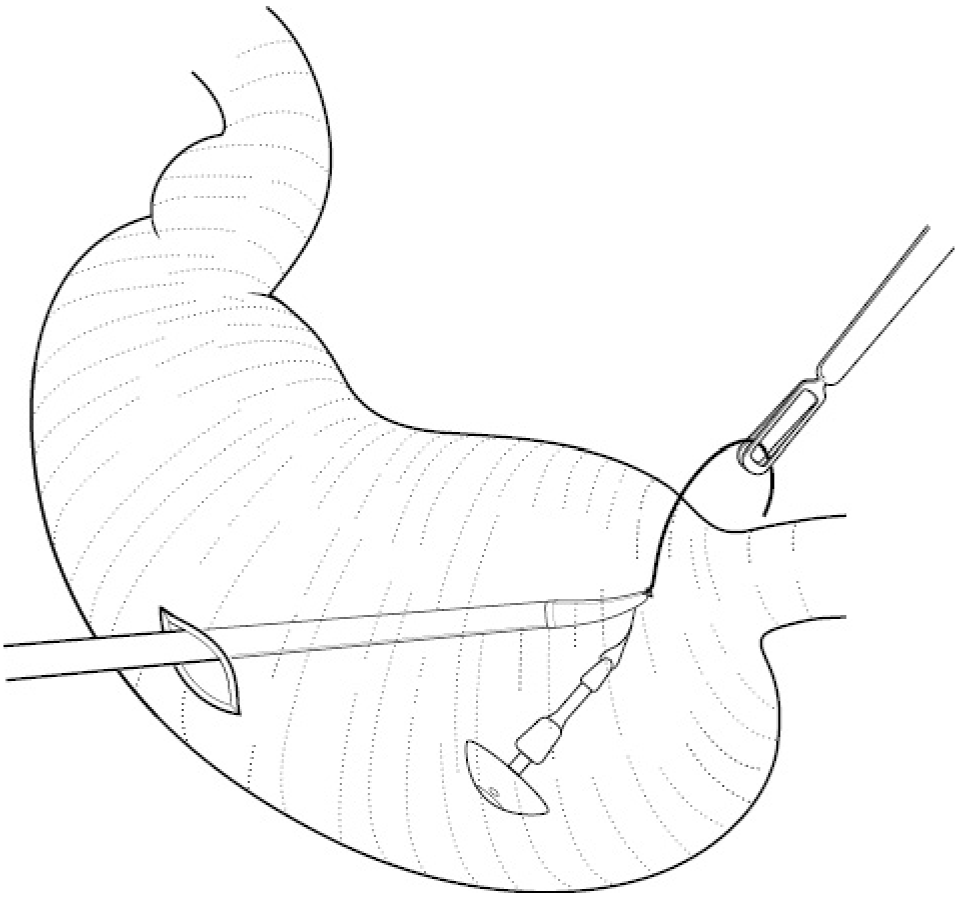
Since the antecolic technique is technically more straightforward and does not risk troublesome bleeding in the transverse mesocolon, it is a favored method by many high-volume bariatric surgeons. In the antecolic technique, the omentum must be draped back inferiorly, and often bivalved up to the transverse colon, to reduce the bulk of tissue that the roux limb will traverse. This omental division also reduces the tension (and thus risk of stricture) transmitted from the Roux limb to the anastomosis when the patient assumes an erect posture postoperatively. Finally, some surgeons have used the bivalved omentum to “wrap” the gastrojejunal anastomosis at the conclusion of the operation. This technique has theoretical benefit in the event of leakage of the gastrojejunal anastomosis, but it is not commonly performed.

Gastric pouch construction

A locking, toothed grasper is next inserted below the xiphoid, applied to tissue just superior to the right crus of the diaphragm, and used to retract the left lobe of the liver anteriorly to expose the gastroesophageal junction. The lesser curve gastric pouch will be created beginning with the dissection at the angle of His. A sufficient plane is then created between the stomach and diaphragm at the angle of His, extending to the left crus, utilizing dissection both bluntly and with the Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA).

Next, a small gastrotomy is made along the greater curvature of the stomach with the Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA). The left upper quadrant (LUQ) 12 mm port site is then enlarged with a Kelly hemostat to allow passage of the 25 mm EEA stapler. The plastic tip of the anvil is dulled gently by the scrub assistant and a loop of Vicryl□ suture is inserted through its end prior to its insertion. The anvil is then passed through the enlarged fascial defect in the LUQ and into the abdomen. Often, a 15 mm XCEL□ trocar is needed to be placed within this newly created fascial defect to allow for adequate insufflations for the remainder of the procedure. The Vicryl□ loop is then grasped with the articulating grasper and the entire anvil is placed through the previously made gastrotomy site (**Figure 33.3**). A site is chosen approximately 3–4 cm inferior to the esophageal hiatus along the lesser curvature of the stomach for the proposed gastrojejunostomy site. The point of the articulating grasper is then brought out through this proposed site with the aid of the

Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA). Once this is accomplished, the suture is grasped and the connecting end (with the plastic insert) is brought out in its entirety through this site. The gastrotomy is then closed utilizing one or two loads from the blue stapler.

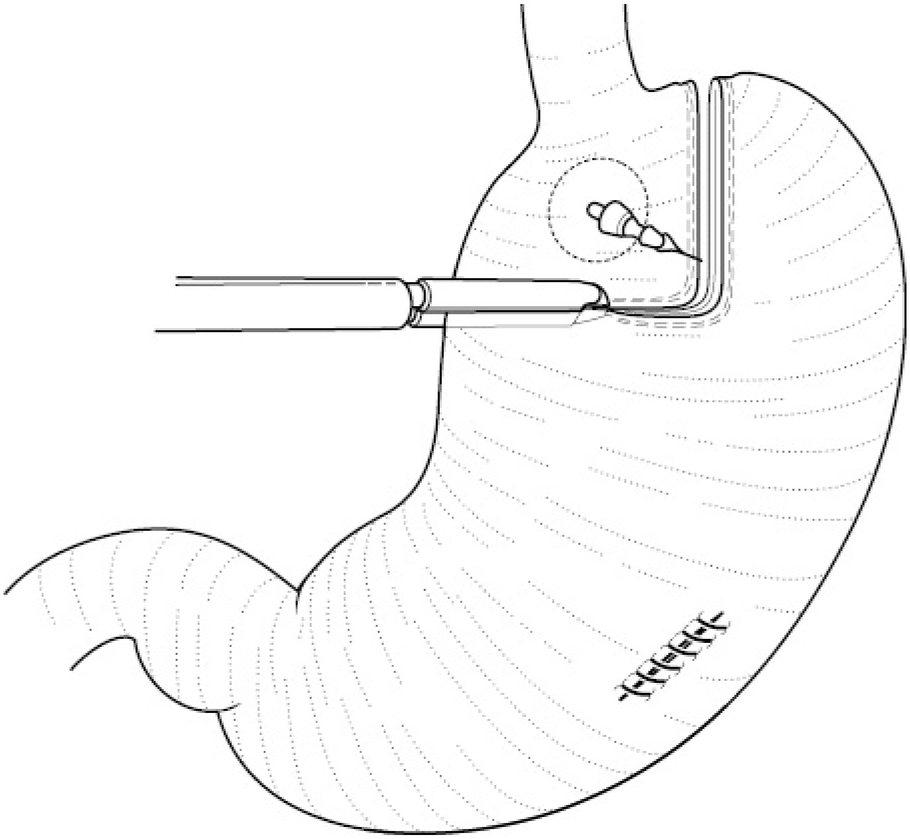


33.3

The creation of the gastric pouch begins with perigastric Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA) dissection along the lesser curvature approximately 6–10 cm inferior to the gastroesophageal junction (usually just below the second major lesser curve vessel). The “flat” side of the blade is used to ensure adequate hemostasis during this dissection. Bipolar electrocautery is also commonly needed. This dissection is continued posteriorly in close proximity to the gastric wall until the plane nearly reaches the lesser sac behind the stomach.

The greater curvature of the stomach is next elevated and a thin area of the gastrocolic ligament is chosen for entry into the lesser sac, using the Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA). Numerous posterior attachments between the lesser curve and retroperitoneum are divided to achieve continuity between the lesser curve dissection, which was performed anteriorly.

The dissection is continued gently such that the tip of the Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA) traverses the posterior surgical plane to emerge anteriorly when the stomach is released. A tubular lesser curve pouch is next created using the Echelon 60 mm stapler with multiple blue loads. The first transverse cut across the lesser curvature is achieved by abutting against the anvil of the EEA stapler, traversing the lesser curve dissection plane created above (**Figure 33.4**). Once in place across the lesser curve, the stapler is fired, creating the inferior-most margin of the pouch. Next, gentle dissection is performed at the apex of the staple line to completely break into the retrogastric lesser sac from above. This simple maneuver is critical since the procedure is far less difficult if the lesser sac is identified and entered from the anterior aspect at this point. The vertical staple line is advanced sequentially by multiple applications of the Echelon 60 mm stapler along the anvil of the EEA stapler. Once the angle of His is reached, the pouch is complete.

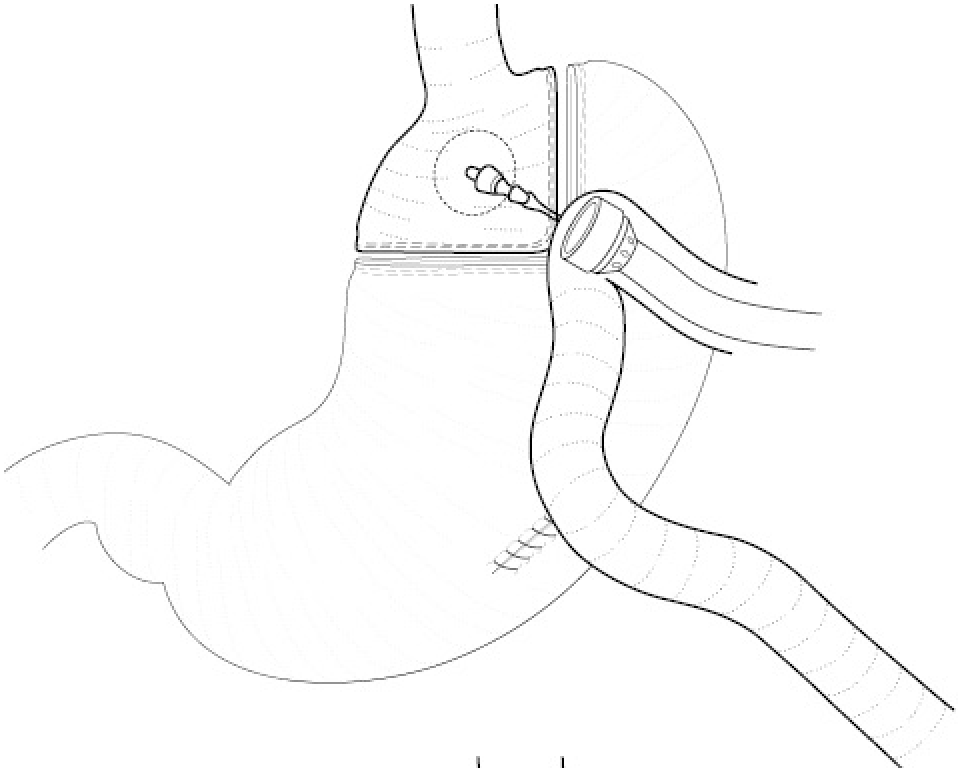


33.4

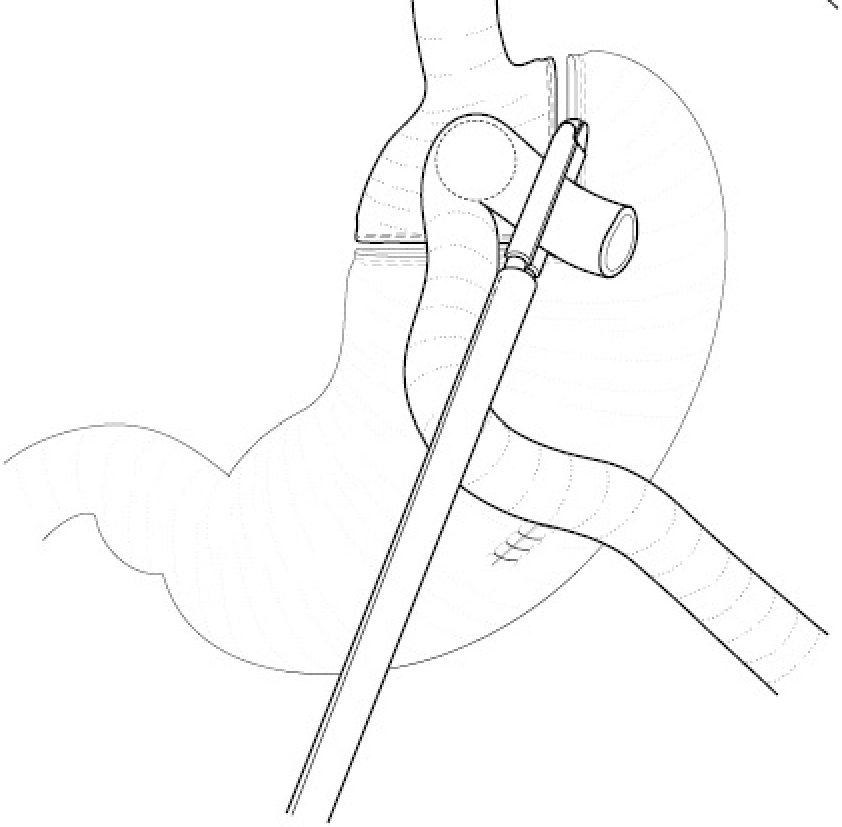
Gastrojejunal anastomosis

There are numerous techniques for laparoscopic gastrojejunostomy including handsewn, the end-to-end stapled technique with the anvil inserted orally (akin to the percutaneous endoscopic gastrostomy (PEG) method of gastrostomy), a linearly stapled technique, and the end-to-end stapled technique with anvil inserted into the pouch laparoscopically. When considering choices of technique, it is usually best to learn from someone who will be readily available for technical consultation, and adopt the technique that he or she employs. The remainder of this chapter will focus on details and pearls for performance of the EEA-stapled gastrojejunostomy technique.

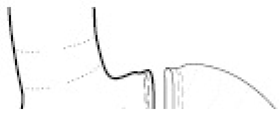
With the gastric pouch created and the anvil of the EEA stapler in place and in full view, the previously fashioned Roux limb is brought up to the gastric pouch in an antecolic fashion. The end of the Roux limb is then opened along the antimesenteric border for approximately 4–6 cm with the Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA), enough space to accept the end of the 25 mm EEA stapler head. The EEA stapler is loaded with a Gore staple-line reinforcement insert and is introduced through the previously widened LUQ port. To protect the Gore staple-line reinforcement insert, a plastic sheath is placed over the stapler. This portion of the procedure can be tedious unless the port site has been widened appropriately. Once the stapler has been inserted, the plastic sheath is pulled away and the end of the EEA stapler is inserted into the open end of the Roux limb. Counter-traction is necessary to ensure that the entire stapler head is within the lumen. With continued traction, the stapler end is slowly opened until the internal mating device pierces through the small bowel (**Figure 33.5a**). The pin is then removed from the end of the anvil. The anvil grasper is then used to mate the two ends of the stapler together. Gentle traction is applied below the proposed anastomosis to ensure that no stray tissue is within the final anastomosis. The stapler is closed, fired, and removed from the port site with the anvil intact. One should check to ensure that two separate donuts of tissue are contained within the stapler complex at its inspection. The 15 mm trocar is then replaced to allow for adequate insufflation. The Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA) is used to take down the mesentery along the mesenteric border of the remaining section of the Roux limb lateral to the anastomosis (**Figure 33.5b and c**). An Echelon 60 mm stapler with white load is used to close the enterotomy, thus completing the anastomosis.

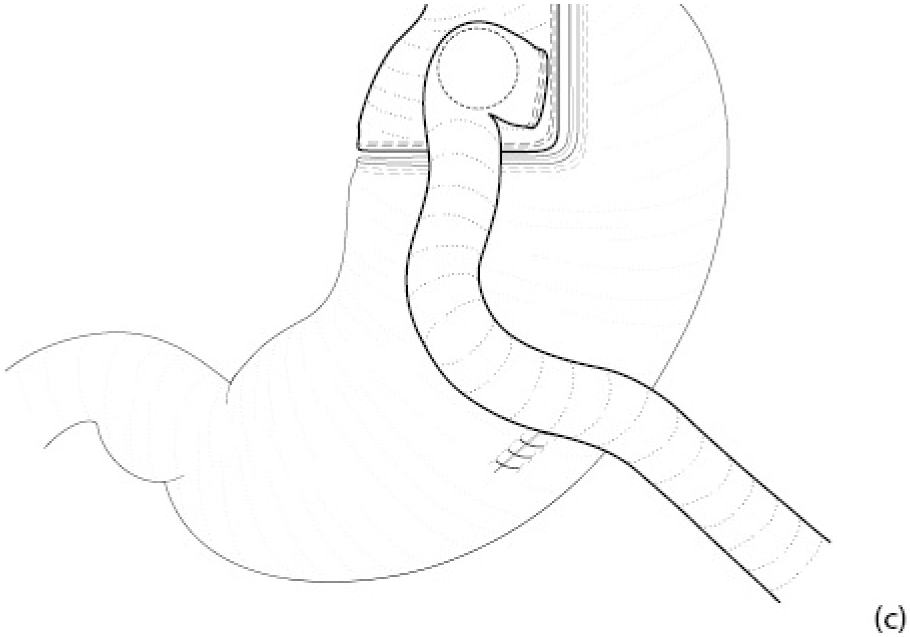


(a)



(b)





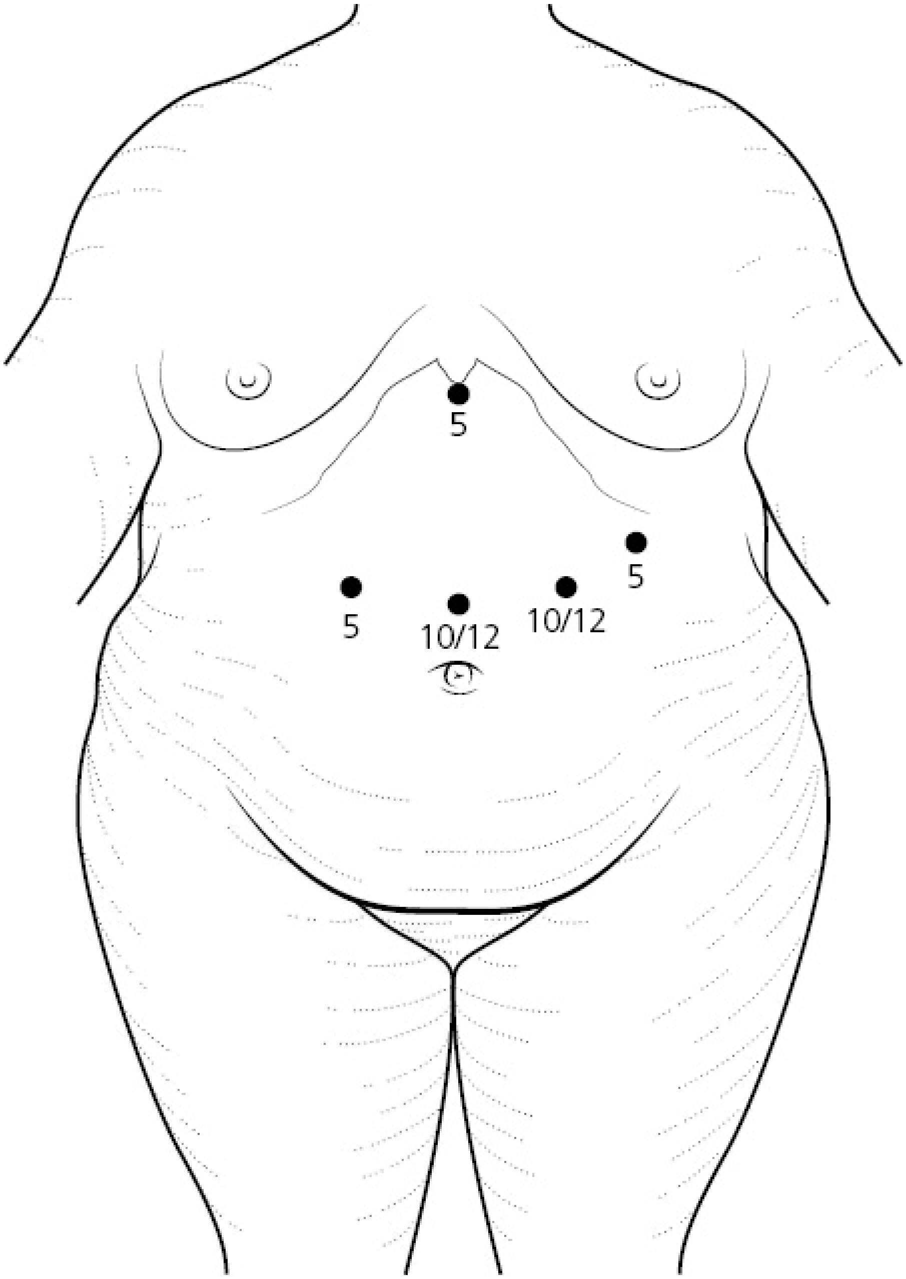
33.5a–c

The integrity and patency of the anastomosis is assessed laparoscopically with intraluminal air insufflation under saline. This pneumatic test is accomplished by temporarily obstructing the Roux limb just beyond the anastomosis and having the anesthesiologist rapidly bolus 60 mL of air into an orogastric tube.

A drain to prevent fluid collection within the abdomen and to provide for early identification of a leak is left near the anastomosis and exits via the patient's upper right trocar site. A temporary gastrostomy tube can be placed in the remnant stomach body if any intraoperative technical challenges are deemed significant enough to warrant deliberate decompression of the bypassed gastric remnant. With the use of the bladeless trocars, the 5 mm and 12 mm port sites do not require fascial closure. The 15 mm port site is closed utilizing the Carter-Thomason CloseSure System in a figure-of-eight type fashion with zero Vicryl® suture. The remaining skin incisions are closed in the standard surgical fashion.

Laparoscopic sleeve gastrectomy operative procedure

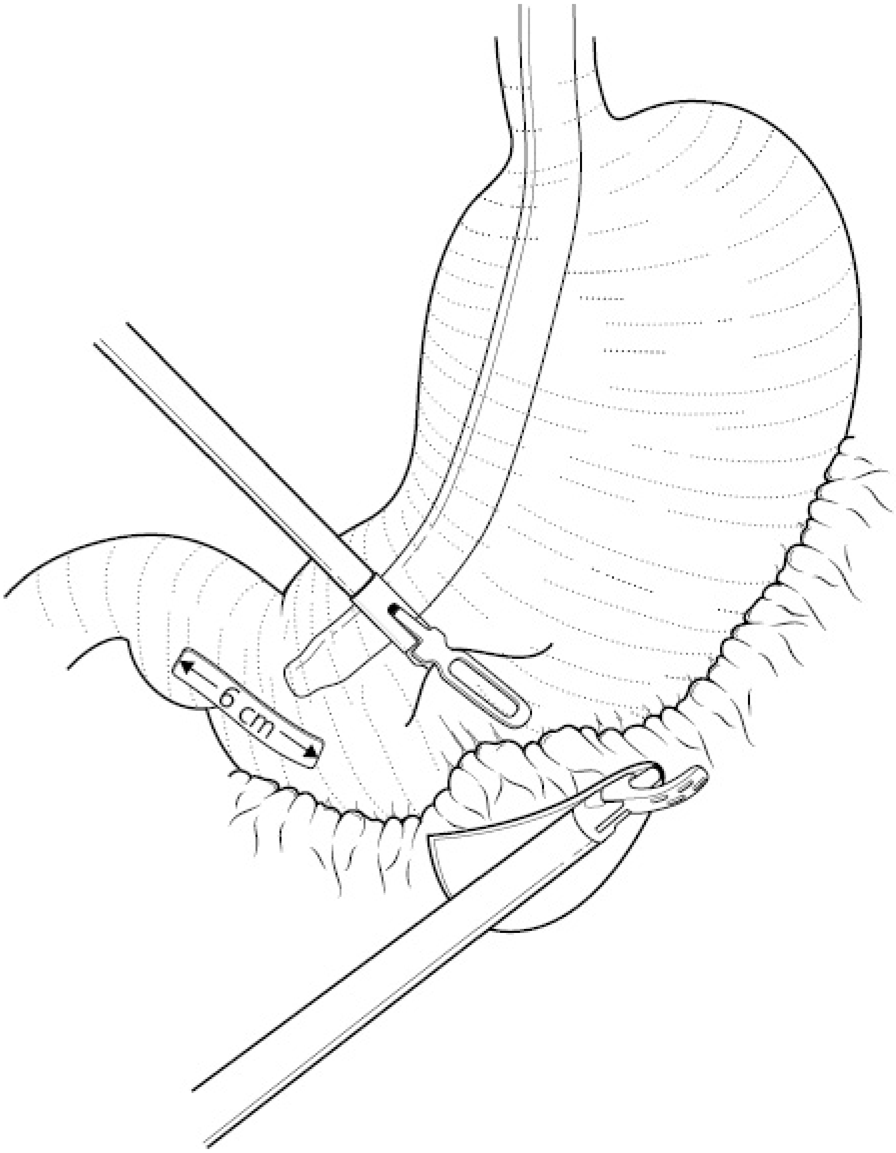
The patient is positioned in a similar manner as previously described for the RYGBP procedure. Two 12 mm and three 5 mm trocars are utilized for the procedure and placed in a similar manner as previously described (**Figure 33.6**). A 34 Fr orogastric tube is placed by anesthesia at the beginning of the case and placed on suction to aid in the dissection. The liver retractor is placed in a similar manner as previously described.



33.6

Greater curvature dissection

An umbilical tape, cut to 6 cm in length, is placed within the abdomen and used to measure the distance from the distal extent of the pylorus along the greater curvature. This is the point where the dissection and initial resection will begin. The greater omentum is then carefully dissected away from the greater curvature of the stomach at this point until the lesser sac is entered. This dissection is carried to the level of the left crus of the diaphragm (**Figure 33.7**). The 34 Fr orogastric tube aids in the identification of the esophagus and the left crus during this portion of the procedure. The assistant is invaluable during this dissection, providing traction and counter-traction, given the often large size of the stomach in these cases. All attachments of the fundic portion of the stomach are taken down, as this can sometimes be folded under the body of the stomach at its superior extent.

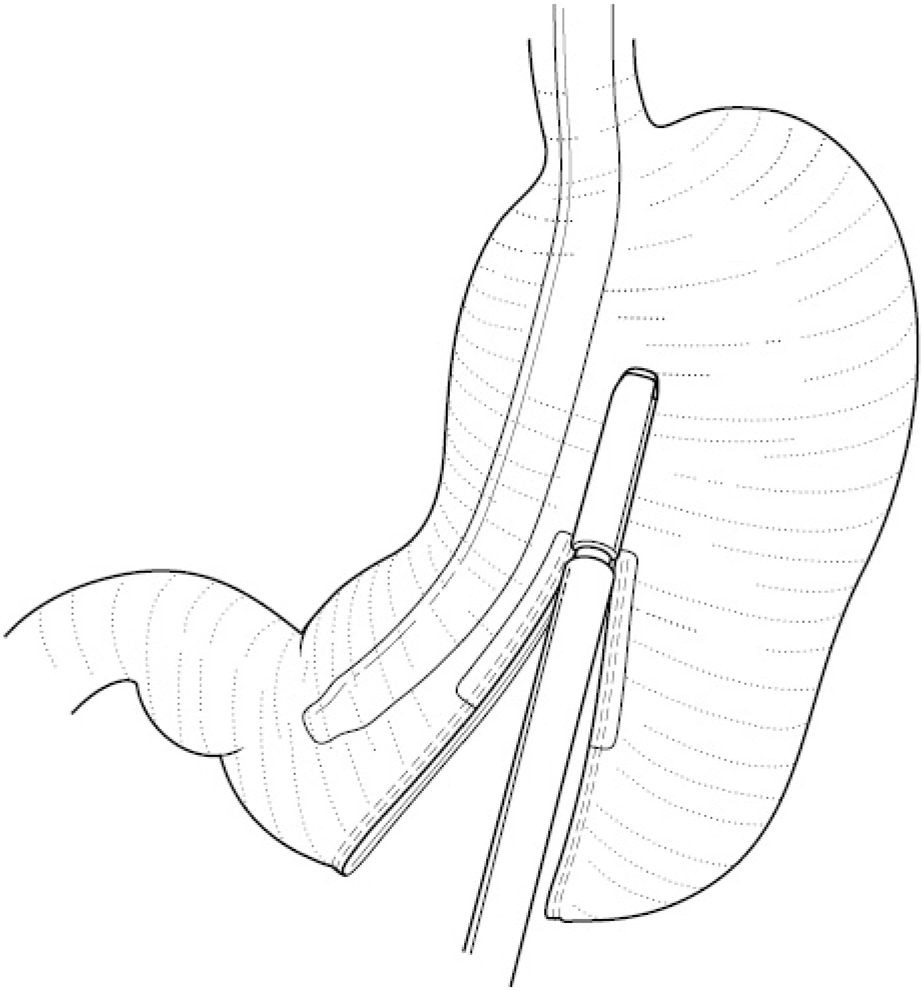


33.7

Gastric resection

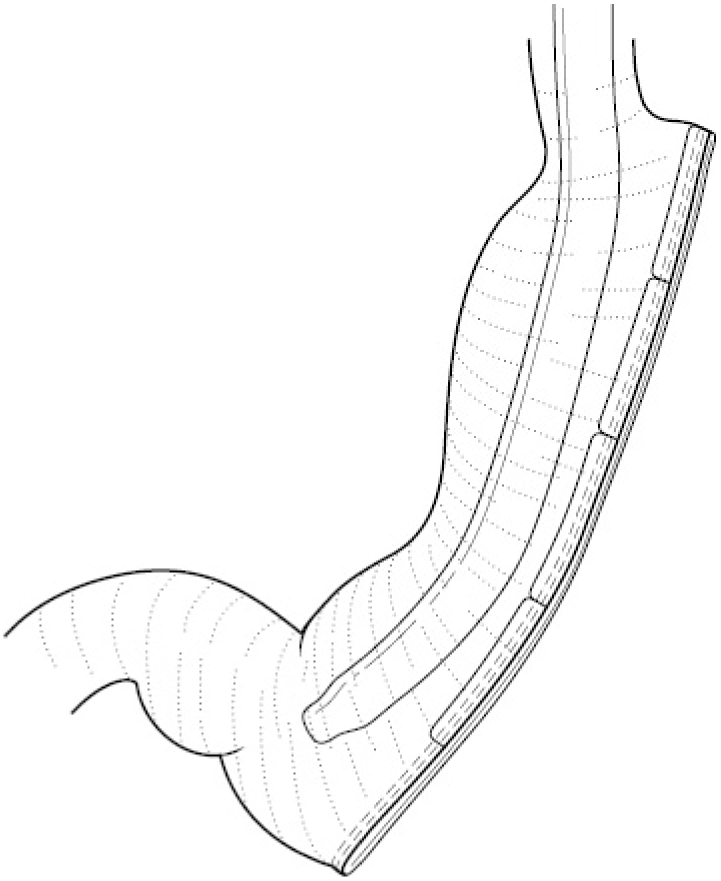
The resectional portion of the procedure is begun by once again measuring from the pylorus to the greater curvature/antrum of the stomach with the 6 cm of umbilical tape. The 34 Fr orogastric tube is then positioned into the antrum and along the lesser curvature of the stomach to act as a guide for the remainder of the procedure. Given its relative thickness, the Echelon 60 mm stapler with a green load, without Gore Seamguard® Bioabsorbable

Staple Line Reinforcement, is used for the first fire across the antrum of the stomach abutting, against the 34 Fr orogastric tube (**Figure 33.8**).



33.8

The remainder of the stapler fires are carried out sequentially along the 34 Fr orogastric tube (adjacent to the lesser curvature of the stomach) with the Echelon 60mm stapler, blue loads, with Gore Seamguard□ Bioabsorbable Staple Line Reinforcement. As the esophagus is approached during the final stages of the resection, it is prudent to veer off slightly lateral to avoid its injury. This does leave a small triangular portion of stomach at the superior aspect of the resection margin, but ensures that the esophagus remains free from the staple line (**Figure 33.9**). This maneuver could potentially decrease the incidence of leaks at this site given the relative proximity of the thin esophageal tissue. The body and fundic portion of the stomach is now free and is removed from the abdomen by gently widening the LUQ 12 mm port site with a Kelly hemostat. The remnant can be placed in a laparoscopic retrieval bag prior to its removal, but, in the authors' experience, we find that the remnant can be removed by simple traction with multiple Kelly hemostats through the widened port site alone.



33.9

The staple line should be tested and the enlarged LUQ 12 mm port is closed in a similar manner as described previously with a drain placed along its length. The skin incisions are once again closed in the standard surgical fashion.

POSTOPERATIVE CARE

Postoperatively, the patients are typically extubated in the operating room after transfer to a hospital bed. They are cared for in a monitored, non-intensive care unit setting, and maintenance fluids are administered based upon lean body weight (typically 40–50% of actual weight). Early warning signs of complication include fever, tachycardia, tachypnea, increasing oxygen requirement, oliguria, hiccoughs, regurgitation, left shoulder pain, worsening abdominal pain, a feeling of anxiety, and acute alteration in mental status. These signs warrant aggressive attention and appropriate investigation since they may signal gastrointestinal leak (anastomosis or staple lines), pulmonary embolus, bowel obstruction, or acute dilation and impending rupture of the bypassed gastric remnant. Routinely, a water-soluble upper gastrointestinal (UGI) contrast study is obtained on postoperative day 1. After satisfactory passage of contrast is documented, patients are begun on clear liquids and subsequently advanced to a high protein liquid diet.

Postoperative monitoring

Bariatric surgery essentially results in surgically enforced very low-calorie, low-carbohydrate dietary intake, thus requiring attention to an adequate intake of important macro- and micronutrients postoperatively. Postoperative follow-up after RYGBP and SG in adolescents is intensive: 2 weeks postoperative visit, then at 6 weeks, 3 months, 6 months, 9 months, 12 months, 18 months, 24 months, then yearly thereafter. Serum chemistries, complete blood count, urine specific gravity, prothrombin time (evidence of vitamin K adequacy) and representative B complex vitamin levels (e.g. B1, B12, folate) are obtained at 6 and 12 months postoperatively then yearly.

Body composition can be assessed with either bioelectrical impedance or dual energy X-ray absorptiometry analysis (DEXA). DEXA not only allows for the measurement of rate and relative amounts of fat and lean body mass loss, but also provides a quantitative assessment of bone mineral density changes. This body composition analysis can be used to modify dietary plans intended to preserve lean body mass during the period of dramatic weight loss. Pregnancy prevention in females is also an important consideration after bariatric surgery due to risks related to rapid weight loss and possible micronutrient deficiencies.

Postoperative diet and medications

Initially, the diet is essentially a protein-rich liquid diet. Dietary advancement after the first month is a methodical process of introducing new items of gradually increasing complexity, toward the goal of a well-balanced, small portion (approximately one cup per meal) diet which ensures a daily intake of 0.5–1 g of protein per kg of ideal weight. Non-steroidal anti-inflammatory medications should be avoided to reduce the risk of intestinal ulceration and bleeding most commonly seen in RYGBP. Ranitidine is prescribed for 6 months postoperatively with some SG patients requiring the transition to a proton-pump inhibitor due to the increased incidence of reflux-type symptoms. Postoperative vitamin and mineral supplementation typically consists of two pediatric chewable multivitamins, a calcium supplement, and an iron supplement for menstruating females. B-complex vitamins are supplemented beyond that which is contained in multivitamin preparations, primarily to augment thiamine and folate supplementation, due to severe complications if deficiency develops.

Five basic “rules” are routinely emphasized with patients and family at each visit:

- Eat protein first.
- Drink 2–3 litres of water or sugar-free liquids daily.
- No snacking between meals.
- Exercise at least 30 minutes per day.
- Always remember vitamins and minerals.

COMPLICATIONS

Table 33.1 summarizes possible complications and their incidence following bariatric surgery.

Table 33.1 **Possible complications following bariatric surgery**

Complication	Percentage (%)	Notes
Gastrointestinal leak requiring reoperation	2–7	Tachycardia is an early warning sign
Gastrojejunal stricture	4–10	Manage with endoscopic dilation
Small bowel obstruction	1–2	High clinical suspicion in RYGB due to increased risk for internal hernias at mesenteric defects
Marginal ulcer	5–10	Treat with PPI; avoid NSAIDS
Bleeding requiring transfusion	1–2	
Wound infection	2–3	Greater with open surgery
DVT/PE	0.5–1	Reduce risk by using mechanical and pharmacologic prophylaxis
Symptomatic cholelithiasis with need for cholecystectomy	8–20	Postoperative rapid weight loss is the main risk factor for cholelithiasis
Nutritional deficiencies	60–70	At risk due to decreased volume and malabsorption (RYGB) as well as poor compliance with taking supplements
Anemia	30–45	Bypass of duodenum (site of iron absorption) contributes to anemia
B12 deficiency	10–20	Peripheral neuropathy is the main symptom, due to decreased intrinsic factor

DVT, deep vein thrombosis; PE, pulmonary embolism.

OUTCOME

Patient satisfaction has generally been very good with this surgical and postoperative approach. The RYGBP and SG operation results in a loss of 60–80% of excess weight over the first year postoperatively. When careful attention is given to adequate protein intake, significant improvements in body composition can be expected after operation. Body composition significantly improves over time, with mean percentage fat mass decreasing from 47% preoperatively to 35% at 12 months. Fat loss appears to plateau by 12 months. This suggests that adolescents undergoing laparoscopic RYGBP dramatically and preferentially reduce body fat mass compared to more modest loss of lean body mass. With regard to comorbidities, both bariatric procedures result in improvement and/or resolution of diabetes, hypertension, hyperlipidemia, and renal function. Lastly, surgery also benefits the patient in terms of psychosocial functioning and quality of life.

VIDEO

OPS_8e_video_33.1 Vertical sleeve gastrectomy

FURTHER READING

Colquitt JL, Picot J, Loveman E, Clegg AJ. Surgery for obesity. *Cochrane Database Syst Rev* 2009; 15: CD003641.

DeLaet D, Schauer D. Obesity in adults. *Clin Evid* 2011; 3: 604.

Inge TH, Courcoulas AP, Jenkins TM, et al. Weight loss and health status 3 years after bariatric surgery in adolescents. *N Engl J Med* 2016; 374: 113–23.

Inge TH, Jenkins TM, Xanthakos SA, et al. Long-term outcomes of bariatric surgery in adolescents with severe obesity (FABS-5+): A prospective follow-up analysis. *Lancet Diabetes Endocrinol* 2017; 5:165–73.

Inge TH, Jenkins TM, Zeller M, et al. Baseline BMI is a strong predictor of nadir BMI after adolescent gastric bypass. *J Pediatr* 2010; 156: 103–8.

Inge TH, Miyano G, Bean J, et al. Reversal of type 2 diabetes mellitus and improvements in cardiovascular risk factors after surgical weight loss in adolescents. *Pediatrics* 2009; 123: 214–22.

O'Brien PE, Sawyer SM, Laurie C, et al. Laparoscopic adjustable gastric banding in severely obese adolescents: A randomized trial. *J Am Med Assoc* 2010; 303: 519–26.

Pratt JS, Lenders CM, Dionne EA, et al. Best practice updates for pediatric/adolescent weight loss surgery. *Obesity* 2009; 17: 901–10.

34

Duodenal atresia and stenosis

AFIF N. KULAYLAT, COLIN G. DELONG, SIMON CLARKE, AND ROBERT E. CILLEY

HISTORY

Successful repair of congenital duodenal obstruction dates back over a century, though description of the pathology was reported as early as 1733 by Calder. Surgical correction was initially made difficult by a lack of appropriate suture material and the lack of understanding of the perioperative care needs of neonates. Improved perioperative care of the sick neonate resulted in many survivors. Historically, three distinct anatomic repairs have been described in the literature, chronologically, the duodenojejunostomy (1916), the gastrojejunostomy (1924), and the duodenoduodenostomy (1943). The gastrojejunostomy was ultimately abandoned secondary to the complications associated with chronic reflux of bilious and pancreatic secretions. By the middle of the 20th century, the duodenojejunostomy, typically performed in a retrocolic, side-to-side fashion, became the standard of care. More recently, duodenoduodenostomy, and its modification the "diamond duodenoduodenostomy" as described by Kimura in 1977, has been used to bypass congenital duodenal obstruction in an effort to hasten the return of intestinal function after surgery and to promote duodenal emptying. As expertise in advanced laparoscopy grows and innovations in technology afford greater versatility, minimal access techniques for treating congenital duodenal obstruction have been demonstrated as safe and feasible.

PRINCIPLES AND JUSTIFICATION

The choice of surgical procedure for the treatment of duodenal obstruction is largely based on the preference of the surgeon. The patient's size or unusual anatomic variants may make one procedure obviously preferred at the time of surgery. Each surgeon should be familiar with the options. The diamond duodenoduodenostomy may result in earlier postoperative feeding and shorter duration of hospitalization. The procedure may be performed using open or laparoscopic surgical techniques. The duodenoduodenostomy is applicable to almost all cases of duodenal obstruction, whether caused by stenosis or atresia, with or without annular pancreas. It may also be used as a safe choice for the treatment of a duodenal web, since it does not risk damage to the pancreaticobiliary system, which may occur with web excision. In the hands of those experienced with the technique, almost any atresia is readily handled using the mobility afforded by the dilated proximal duodenum in combination with mobilization of the distal duodenum beneath the superior mesenteric vessels. A one-layer anastomotic technique is used most often. Although a decompressing gastrostomy was placed at the time of surgery in the past, this is no longer performed routinely. However, in a child with multiple anomalies, in whom poor feeding is predictable, addition of a gastrostomy during duodenoduodenostomy may be beneficial. Laparoscopic duodenoduodenostomy has been shown to be both safe and effective in experienced hands. Potential advantages include earlier postoperative feeding, less pain, and shorter hospitalization. Advances in small articulating instruments may enhance the ability of pediatric surgeons to perform this procedure. The only contraindication to surgery is the presence of multiple severe anomalies incompatible with life.

PREOPERATIVE

Duodenal obstruction may be diagnosed prenatally by fetal ultrasound or fetal magnetic resonance imaging (MRI). A dilated stomach and duodenum are seen on imaging studies. Neonates with congenital duodenal obstruction most often present with obvious symptoms on the first day of life. Feeding intolerance and vomiting, which is usually bilious, are noted from the outset. Vomiting may be non-bilious when preampullary obstruction is present. Dehydration and electrolyte depletion rapidly ensue if the condition is not recognized and IV therapy begun. Parenteral nutrition is frequently initiated as feeding is often delayed after repair. Secondary complications, such as aspiration and respiratory failure, may also be present. The presence of a “double bubble” on a plain abdominal radiograph is essentially diagnostic of duodenal atresia. Air may be seen distally in the gastrointestinal tract with incomplete obstructions (e.g. stenosis) or an unusual double ampulla that opens both above and below the stenosis. Contrast radiography is confirmatory and may be especially helpful in confirming the diagnosis. Differentiating intrinsic duodenal obstruction from malrotation with volvulus is also facilitated with contrast radiography. Duodenal obstruction is treated less urgently than malrotation by some surgeons and therefore differentiating between the two entities is critical. Assuming that the diagnosis of malrotation with midgut volvulus has been excluded, workup and evaluation of other potential associated anomalies may proceed.

Trisomy 21, occurring in 25–40% of affected infants, and congenital heart disease must be suspected in all children with duodenal atresia. Duodenal atresia also occurs as part of the Feingold syndrome and Joubert syndrome and has been reported in Goldenhar syndrome and 47XXX. Concurrent malrotation and second intestinal atresias are gastrointestinal abnormalities that occur with increased frequency in patients with duodenal atresia. The utility of demonstrating distal bowel continuity is debated. Rarely, duodenal atresia has occurred in association with esophageal atresia, hereditary multiple atresias, and developmental abnormalities of the bile ducts (including choledochal cysts and biliary atresia).

ANESTHESIA

General anesthesia with rapid sequence endotracheal intubation is required. Many pediatric anesthetists use epidural anesthetic supplementation for the operation, as well as for postoperative analgesia. Prevention of hypothermia is accomplished by heating the operating room, warming the anesthetic gases, external warming lights, operating table warmers, warming of the IV fluids, and the use of adhesive plastic drapes for surgical draping. Specialized pediatric anesthetic care has been demonstrated to reduce both morbidity and mortality in newborn surgery.

OPERATION

Incision and initial evaluation

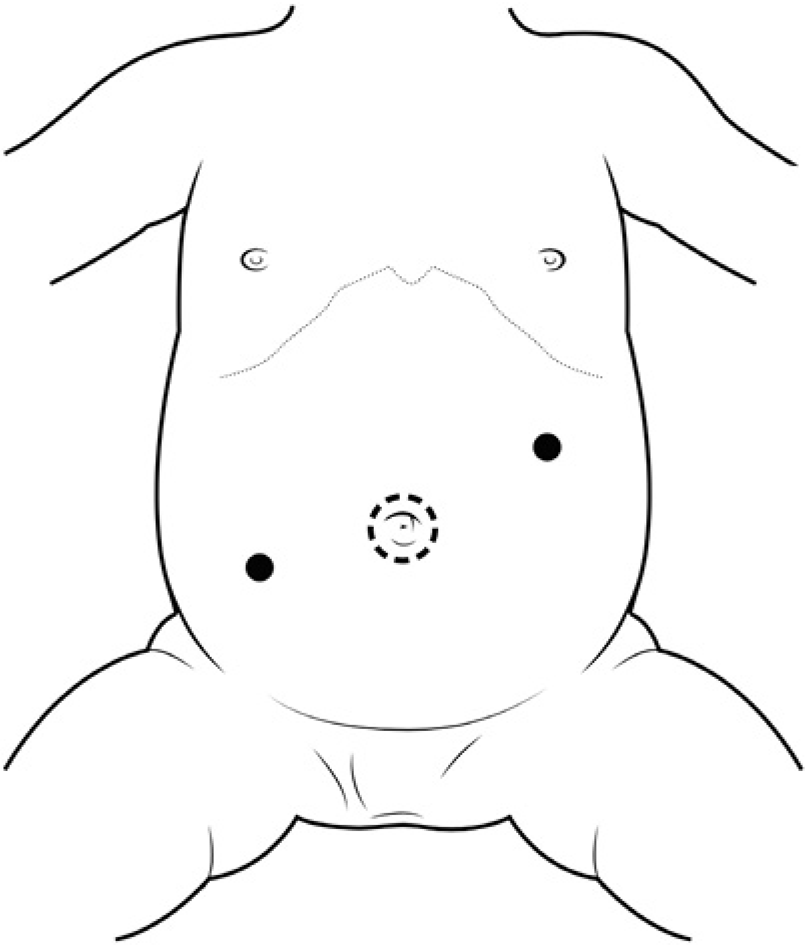
OPEN APPROACH

A small, transverse, right upper quadrant (RUQ) incision provides adequate exposure. Alternatively, a transumbilical laparotomy may be performed. The type and location of the atresia, as well as any pancreatic abnormality or the presence of a rare preduodenal portal vein are noted. The patient is assessed for any abnormality of intestinal rotation; if present, a Ladd's procedure and appendectomy are performed. The presence of a normal gallbladder is noted.

Diagnosis and treatment (timing and sequence) of simultaneous duodenal atresia and esophageal atresia may be difficult. Some surgeons would argue for addressing the tracheoesophageal fistula first, while others would advocate for first addressing the duodenal repair. Simultaneous repair may be appropriate in some patients.

LAPAROSCOPIC APPROACH

The infant is placed in the supine position at the edge of the operating table with the surgeon standing at the infant's feet. The assistant stands on the patient's left. A small infant size umbilical port is placed under direct visualization and secured in place with a purse-string suture to avoid CO_2 loss and minimize port dislodgment. Alternatively, based on surgeon preference and infant size, a Veress needle can be used to access and insufflate the abdomen at the umbilicus, and then exchanged for an infant size port. Some surgeons prefer infraumbilical access and visualization of intraperitoneal structures prior to insufflation to minimize potential for gas embolus through a patent umbilical vein. A 30-degree laparoscope is advisable to facilitate visualization. Insufflation pressures are minimized based on infant size and tolerance. Generally, two additional working ports are placed in the left and right mid abdomen in a staggered fashion relative to the umbilicus, though their positioning can vary based on surgeon preference (**Figure 34.1**). Inspection of the abdomen proceeds as in the open approach.

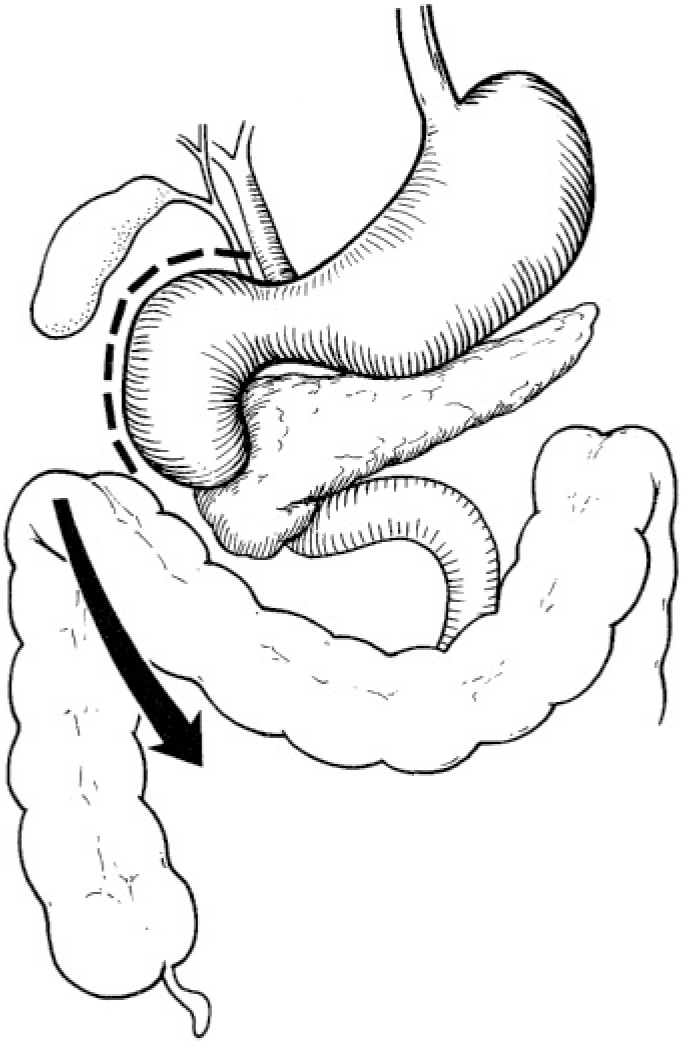


34.1

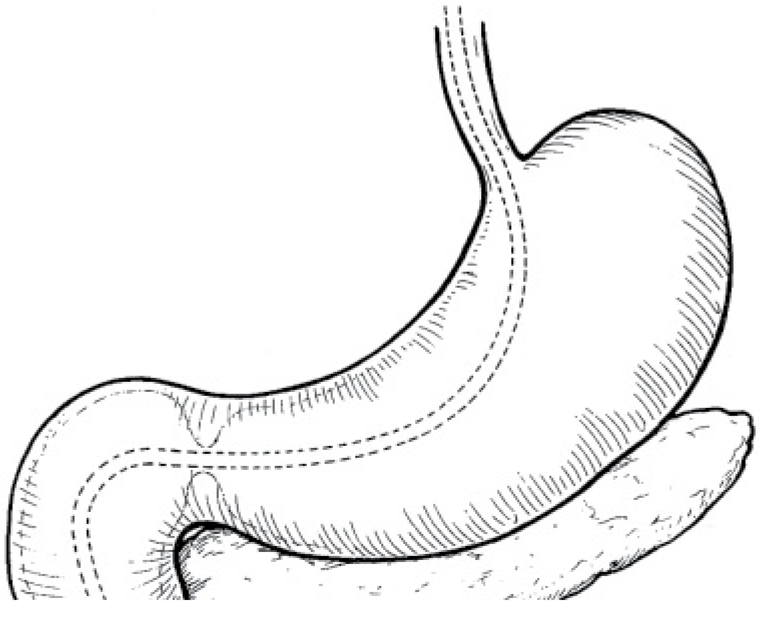
Mobilization and retraction

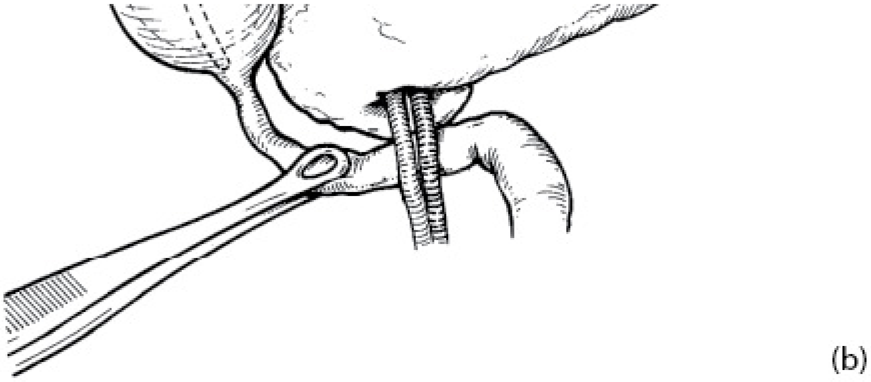
OPEN APPROACH

The hepatic flexure of the colon is mobilized sufficiently to expose the duodenum, and the proximal obstructed duodenum is freed from its retroperitoneal attachments (**Figure 34.2a**). The requirements for distal mobilization vary according to the location of the atresia. If necessary, the entire distal duodenum may be mobilized from beneath the superior mesenteric artery. Rightward traction on the exposed distal duodenum allows these retroperitoneal attachments to be divided. A transpyloric tube may be passed to determine if a "windsock" abnormality is present (**Figure 34.2b**). Either hand-held or fixed, table-mounted retractors may be used. If examination of the proximal segment reveals a grossly dilated duodenum concerning for future dysmotility, consideration for a tapering duodenoplasty is given as it may facilitate return of more functional and effective peristalsis. This is typically accomplished by suture plication or by resection with a stapler. These maneuvers should be performed on the anterior or anterolateral surface to avoid damage to the common bile duct (CBD), pancreas, and ampulla.



(a)



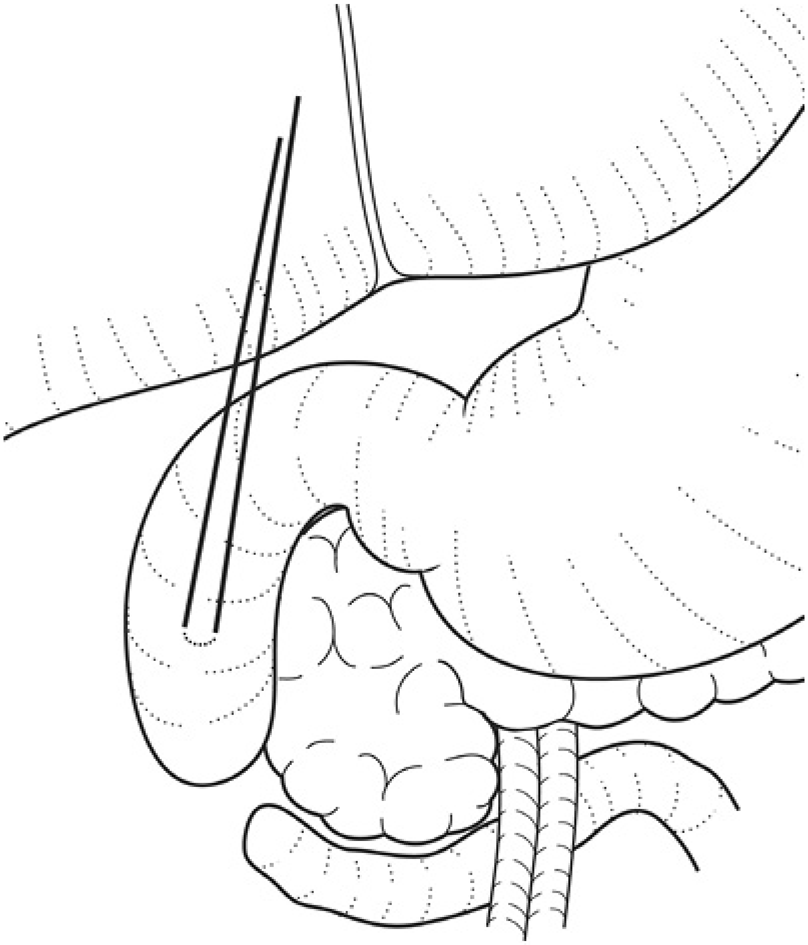


34.2a,b

LAPAROSCOPIC APPROACH

Adequate exposure of the duodenum is essential for success. An additional port may be placed in the left epigastrium or right lower quadrant for elevation/retraction of the liver. Alternatively, a monofilament suture can be taken into the abdomen and out again through the upper abdominal wall gently hitching up the falciform ligament in its path. This is then tied onto the abdominal wall surface over a pledget to avoid marks on the abdominal wall. Laparoscopic kocherization of the duodenum is performed. Care is taken not to injure the pancreas or CBD.

Stay sutures should be placed in the proximal dilated duodenum to stabilize the proximal segment (**Figure 34.3**). Alternatively, some surgeons advocate for a traction suture to be placed through the abdominal wall incorporating both the proximal and distal segment of the duodenum after creation of the duodenotomies to facilitate exposure of the anastomosis for laparoscopic suture placement.

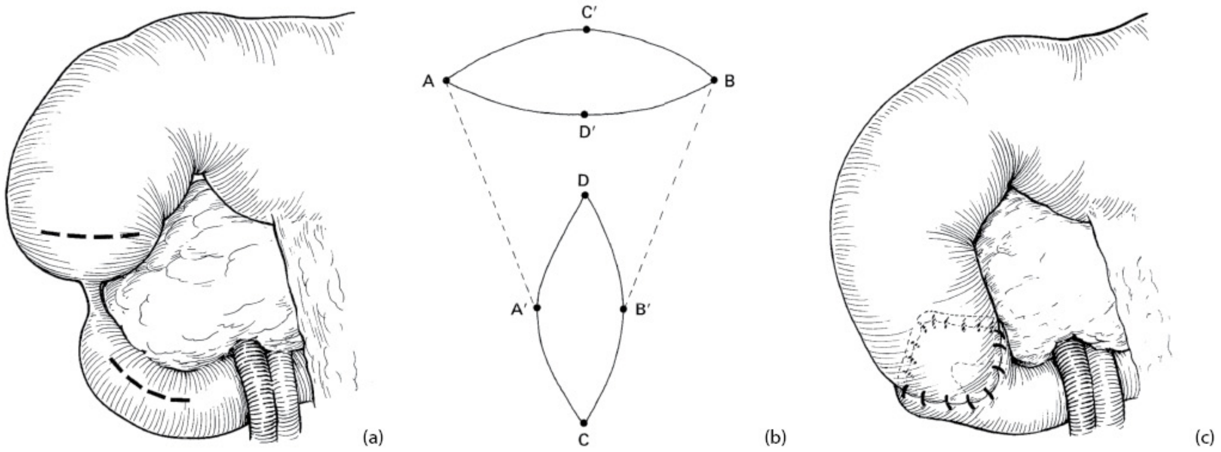


34.3

Duodenoduodenostomy

OPEN APPROACH

A transverse duodenotomy is performed in the proximal segment (**Figure 34.4a**). It is important that this incision be made 1 cm above the atresia to avoid any possibility of injury to the pancreaticobiliary system, which may enter anywhere in the vicinity of the duodenal web, stenosis, or atresia. Retrograde passage of a probe into the stomach confirms the absence of a duodenal web and eliminates the rare possibility of a concurrent gastric antral web. If a web is encountered, excision can be undertaken with cautery, being cautious to avoid the medial aspect of the web, particularly if the ampulla is not well visualized. A longitudinal duodenotomy of the same length is created in the distal segment. The antimesenteric duodenotomies should be 1–2 cm in length based on the size of the patient to ensure an adequately patent anastomosis. Passage of a small tube distally confirms patency of the distal duodenum. Injection of air or saline into the distal segment is conveniently performed at this stage to rule out a second atresia if deemed necessary. Some also utilize this technique with distal occlusion of the bowel to slightly stretch the intestinal wall, arguably facilitating an easier anastomosis. If there is difficulty mobilizing the distal segment, division of the ligament of Treitz may help facilitate an appropriately oriented and tension-free anastomosis. A single layer of interrupted sutures with posterior knots tied inside and anterior knots tied outside ensures symmetry. The orientation of the sutures in the "diamond" anastomosis is shown in **Figure 34.4b** and the completed anastomosis is shown in **Figure 34.4c**. The anastomosis may be performed with permanent or absorbable sutures in an interrupted or continuous fashion.



34.4a–c

If a long atretic segment is encountered, mobilization of the distal duodenum is difficult, or the procedure is challenging due to patient anatomy (particularly in small, premature infants), a duodenojejunostomy may be performed. The first portion of the jejunum beyond the ligament of Treitz is passed through a small fenestration in the transverse colon mesentery for a retrocolic duodenojejunal anastomosis. Recent literature suggests equivalent results for this operation.

A gastrostomy may be performed if the need is anticipated. Otherwise an orogastric tube is confirmed to be in the appropriate position. Some advocate for transanastomotic feeding tubes, though no clear benefit has been consistently demonstrated. The distal bowel is examined for an additional atretic segment though the incidence is reportedly <1%.

LAPAROSCOPIC APPROACHES

Once the transverse incision is made on the proximal segment with hook diathermy, the duodenotomy should be extended with scissors as to avoid possible thermal damage to the ampulla. Laparoscopic suturing technique and intracorporeal knot tying is used to perform the anastomosis in similar fashion to the open approach. Either running sutures or single interrupted sutures can be used for the anterior and posterior walls, at the discretion of the surgeon. As previously discussed, apical stitches can be tied extracorporeally and left on tension through the abdominal wall to help align the enterotomies. Based on the orientation and lay of the proximal and distal segment of duodenum, some surgeons find a traditional side-to-side anastomosis to be technically more feasible when approached laparoscopically. “U-clips” have also been advocated as alternative and safe technique to perform the anastomosis. Though it may be challenging, the surgeon can still similarly evaluate the distal bowel for any obvious atresias, although, as described previously, the incidence has been noted to be <1%. Ports should be removed under direct visualization, to ensure no omental evisceration. All ports are closed. Skin can be closed with tissue glue or suture.

POSTOPERATIVE CARE

Postoperative care consists primarily of supportive measures to provide nutrition while awaiting the return of intestinal function. Immediate enteral feeding can be started if a transanastomotic tube is placed at the time of the initial operation. Transanastomotic feeding may reduce parenteral nutrition use and improve the time to oral feeding. The disadvantages of transanastomotic feeding include tube dislodgment and intestinal injury. Parenteral nutrition may be used to provide nutritional support postoperatively. To minimize the risks of parenteral nutrition (especially hepatotoxicity), total calories, protein, and fat intake should be kept at the lowest levels possible to allow growth. Peripherally inserted central venous catheters are used commonly.

Unless the child was septic before surgery, only a prophylactic course of antibiotics is indicated. Ventilatory support is provided as needed. Nasogastric or gastrostomy drainage is maintained until gastric emptying begins, as heralded by a change in the quality of the gastric drainage from bilious green to clear or yellow and a decrease in gastric residuals. Feeding is instituted slowly and may require a period of several weeks before full enteral nutrition is tolerated. Standard infant formulas or breast milk are satisfactory; partially digested formulas are usually unnecessary. The need and timing for postoperative contrast studies are debated. There is a trend toward earlier attempts at feedings, often dictated by a non-obstructed contrast series.

OUTCOME

The outcome depends primarily on the presence of other anomalies. Survival has increased dramatically with improved anesthetic care and nutritional support. Selective pregnancy termination has also played a role. Almost all mortality is now related to associated anomalies of other organ systems, primarily heart, lungs, and brain. Anastomotic leak, intra-abdominal sepsis, and wound complications occur rarely. Prolonged feeding intolerance is the most common complication. While prokinetic agents have been proposed, none have been found to have a consistent benefit. In the absence of intraoperatively noted issues, return of gastrointestinal function is expected by approximately 3 weeks. Upper gastrointestinal series may be useful to search for residual obstruction, anastomotic stenosis, or unrecognized obstruction at a different location, missed second atresia, missed “windsock” deformity, or poor peristalsis. Duodenal atony or paresis with a functional duodenal obstruction in the face of an anatomically patent anastomosis is a rare but frustrating problem. Plication of the duodenum or an alternative method of duodenal bypass is a surgical option if conservative observation is unsuccessful.

Long-term complications are uncommon. Symptoms such as pain, vomiting, and feelings of fullness may be present in up to one-third of patients when studied as adults. The symptoms correlate poorly with objective findings on upper gastrointestinal radiographic studies and endoscopy. Late non-function may respond to duodenal plication. Long-term follow-up of these patients is recommended.

FURTHER READING

Ein SH, Kim PC, Miller HA. The late nonfunctioning duodenal atresia repair – a second look. *J Pediatr Surg* 2000; 35: 690–1.

Escobar MA, Ladd AP, Grosfeld JL, et al. Duodenal atresia and stenosis: Long-term follow-up over 30 years. *J Pediatr Surg* 2004; 39: 867–71.

Fonkalsrud EW, DeLorimier AA, Hays DM. Congenital atresia and stenosis of the duodenum. *Pediatrics* 1969; 43: 79–83.

Jensen AR, Short SS, Anselmo DM, et al. Laparoscopic versus open treatment of congenital duodenal obstruction: Multicenter short-term outcomes analysis. *J Laparoendosc Adv Surg Tech A* 2013; 23: 876–80.

Kimura K, Mukohara N, Nishijima E, et al. Diamond-shaped anastomosis for duodenal atresia: An experience with 44 patients over 15 years. *J Pediatr Surg* 1990; 25: 977–9.

Rothenberg SS. Laparoscopic duodenoduodenostomy for duodenal obstruction in infants and children. *J Pediatr Surg* 2002; 37: 1088–9.

Spilde TL, St Peter SD, Keckler SJ, et al. Open vs laparoscopic repair of congenital duodenal obstructions: A concurrent series. *J Pediatr Surg* 2008; 43: 1002–5.

St Peter SD, Little DC, Barsness KA, et al. Should we be concerned about jejunoileal atresia during repair of duodenal atresia? *J Laparoendosc Adv Surg Tech A* 2010; 20: 773–5.

Valusek PA, Spilde TL, Tsao K, et al. Laparoscopic duodenal atresia repair using surgical U-clips: A novel technique. *Surg Endosc* 2007; 21: 1023–4.

Weber TR, Lewis JE, Mooney D, Connors R. Duodenal atresia: A comparison of techniques of repair. *J Pediatr Surg* 1986; 21: 1133–6.

Zani A, Yeh JB, King SK, et al. Duodeno-duodenostomy or duodeno-jejunostomy for duodenal atresia: Is one repair better than the other? *Pediatr Surg Int* 2017; 33: 245–8.

35

Malrotation

SIMON BLACKBURN, JOSEPH I. CURRY, AND BHANUMATHI LAKSHMINARAYANAN

INTRODUCTION

The term “malrotation” refers to a condition in which the midgut – that part of the intestine supplied by the superior mesenteric vessels and extending from the duodenojejunal flexure to the mid-transverse colon – remains unfixed and suspended on a narrow-based mesentery.

HISTORY

The first description of intestinal development was written by Mall in 1898. Frazer and Robins in 1915 expanded on the observations of Mall, and in 1923 Dott extended the embryological observations to the problems encountered clinically. In 1936, William Ladd emphasized the importance of releasing the duodenum and placing the cecum in the left upper quadrant (LUQ) and, indeed, the principles of the modern procedure are almost unchanged from those of Ladd.

PRINCIPLES AND JUSTIFICATION

Embryology

The intestines initially develop as a straight tube extending down the midline of the embryo. As it lengthens, it extends into the extra-embryonic celom of the umbilical cord, but later returns to the abdominal cavity. The *foregut (stomach and duodenum)* is supplied by the celiac artery, the *midgut (small intestine and proximal colon)* by the superior mesenteric artery, and the *hindgut (mid-transverse colon to the rectum)* by the inferior mesenteric artery. Three stages of development of the midgut are recognized.

STAGE I

The first stage occurs during weeks 4–10 of gestation. Because of rapid growth, the coelomic cavity is unable to contain the midgut within its confines. The midgut is forced out into the physiologic hernia within the umbilical cord. During this process, the first 90-degree anticlockwise rotation of the midgut is observed.

STAGE II

Stage II occurs at weeks 10–12, during which time the midgut migrates back into the abdomen. The small intestine returns first and lies mainly on the left side of the abdomen. The cecocolic loop returns last, entering the abdomen in the left lower quadrant. A further 90-degree anticlockwise rotation occurs during this phase.

STAGE III

Following the return of the midgut to the abdomen, the final 90 degrees of rotation occurs. During week 12 of gestation, various parts of the mesentery and the posterior parietal peritoneum fuse, notably the cecum and the ascending colon, which become fixed in the right paracolic gutter. The duodenojejunal loop comes to rest behind and to the left of the superior mesenteric vessels. The cecocolic loop lies in front of and to the right of these vessels.

CONSEQUENCES OF ERRORS OF NORMAL ROTATION

Errors may occur at any of the three stages, with varying consequences.

Stage I

Failure of the intestine to return into the abdomen results in the formation of an exomphalos/omphalocele (see Chapter 25).

Stage II

During this stage, a number of errors may occur:

- *Non-rotation:* Rotation may fail to occur following re-entry of the midgut into the abdomen.
- *Incomplete rotation:* Anticlockwise rotation is arrested at 180 degrees. The cecum lies in a subhepatic position in the right hypochondrium. The duodenojejunal rotation is similarly arrested and the duodenojejunal flexure lies to the right of the midline and superior mesenteric vessels. The base of the midgut mesentery is compressed and narrow, and the entire midgut hangs suspended on the superior mesenteric vessels by a narrow stalk, which is prone to volvulus.
- *Reversed rotation:* The final 180-degree rotation occurs in a *clockwise* direction, with the colon coming to lie posterior to the duodenum and superior mesenteric vessels.
- *Hyper-rotation:* Rotation continues 360 degrees or more so that the cecum comes to rest in the region of the splenic flexure in the left hypochondrium.
- *Encapsulated small intestine:* The avascular sac that forms the lining of the extraembryonic celom returns *en masse* into the abdomen with the intestine.

The most common error is incomplete rotation. Treatment of this condition forms the basis of the rest of the discussion.

Stage III

Rotation occurs normally but fixation is defective, resulting in a “mobile cecum.” It is estimated that this situation is present in 10% of asymptomatic individuals, but it may predispose to cecal volvulus or to intussusception.

INCIDENCE

Approximately 60% of clinical cases present in the first month of life, and more than two-thirds of these within the first week. Sporadic cases occur throughout life. It is generally accepted that once the diagnosis of malrotation has been established, surgical correction is mandatory to prevent the development of volvulus, which occurs in 40–50% of cases not treated surgically. Disorders of rotation are associated with exomphalos/omphalocele, gastroschisis, congenital diaphragmatic hernias, and prune-belly syndrome. They are also found in conjunction with intrinsic duodenal obstructions, esophageal atresia, Hirschsprung disease, biliary atresia, congenital heart anomalies, and urinary tract anomalies. Associated anomalies are present in about 30% of patients.

CLINICAL PRESENTATION

Neonatal period

Malrotation in early infancy may present either with acute strangulating obstruction or with recurrent episodes of incomplete intestinal obstruction.

Acute, life-threatening, strangulating intestinal obstruction occurs as a result of midgut volvulus. The infant presents in a shocked and collapsed state with bilious vomiting (which often contains altered blood), abdominal tenderness with or (more commonly) without distension, and the passage of dark blood rectally. Edema and erythema of the abdominal wall develop as the volvulus becomes complicated by intestinal gangrene, perforation, and peritonitis.

Infants and children

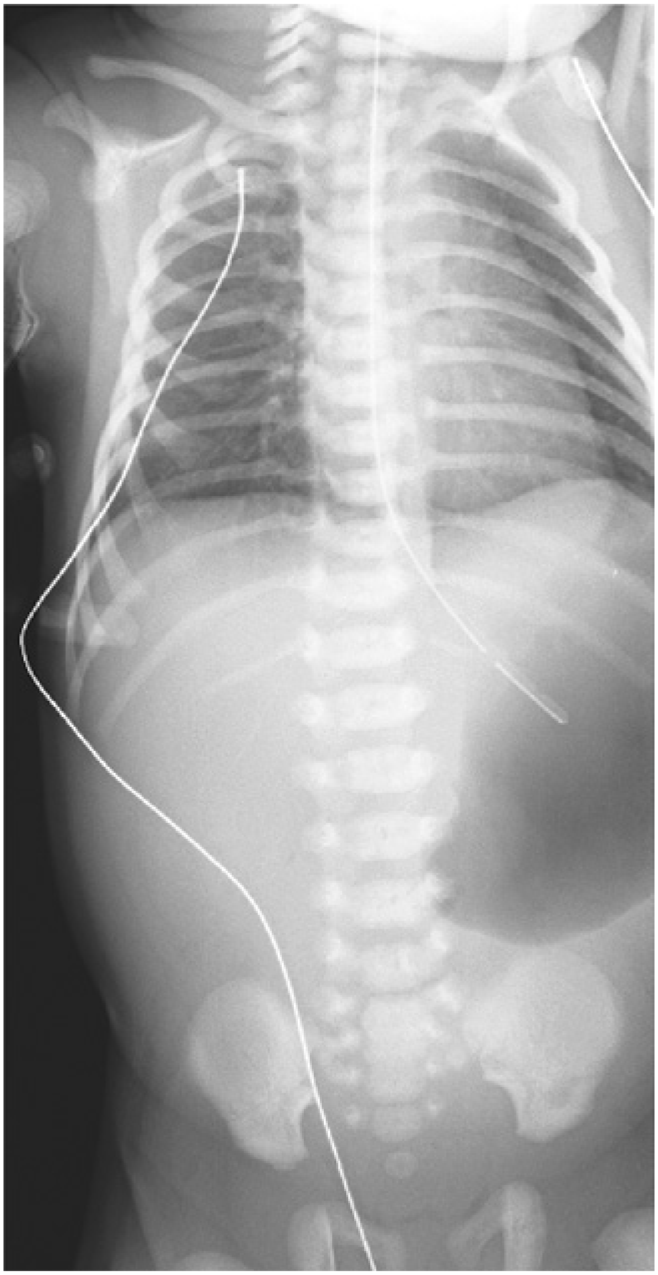
A wide spectrum of clinical symptoms has been ascribed to malrotation. The most common symptom is intermittent vomiting, which is often bile-stained. Failure to thrive and malrotation may be a result of intestinal malabsorption secondary to lymphatic compression in the narrow-based mesentery of the small intestine. Older children may present with features of anorexia nervosa. Early satiety or pain associated with intake of food results in a reluctance to eat or food aversion.

PREOPERATIVE

Radiologic investigations

PLAIN ABDOMINAL RADIOGRAPHY

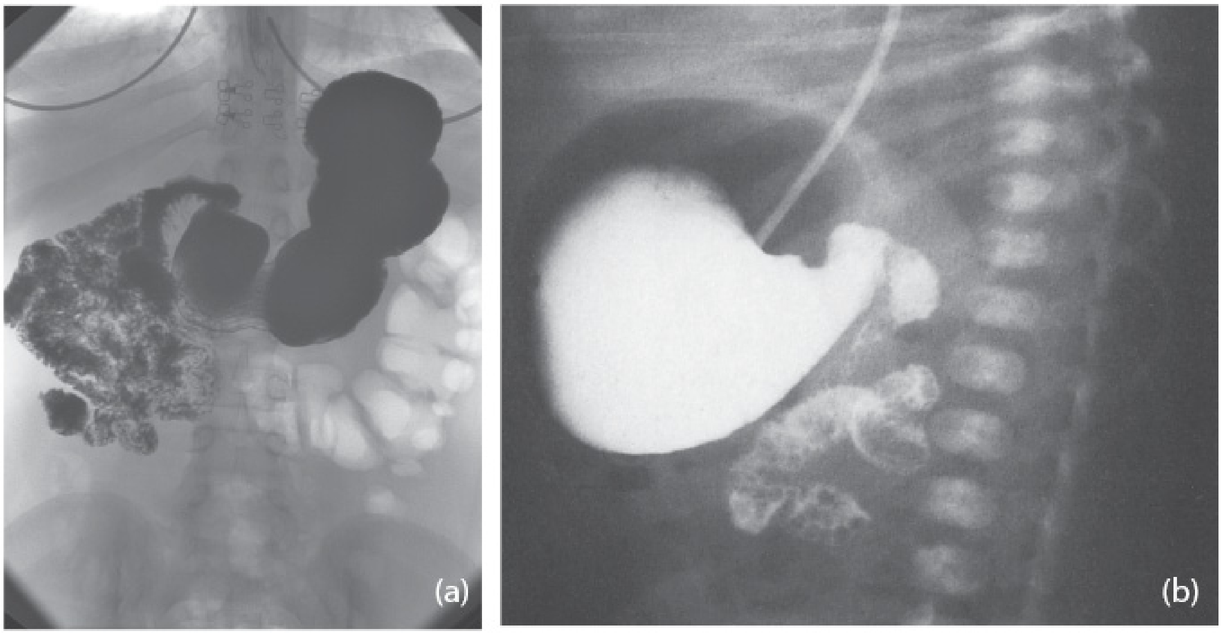
The features suggestive of a volvulus on the plain abdominal radiograph are an air-filled stomach and a paucity of gas in the rest of the intestine (**Figure 35.1**). The abdominal X-ray may, however, be normal. In the infant presenting in shock with features of acute strangulating obstruction, further radiologic investigations only delay definitive treatment.



35.1

CONTRAST RADIOLOGY

The investigation of choice is an upper gastrointestinal contrast study. The features that should be elicited are an abnormal configuration of the duodenal C-loop, the identification of the duodenojejunal flexure to the right of the midline, and small bowel loops on the right side of the abdomen (**Figure 35.2a and b**). A "twisted ribbon" and "corkscrew" appearance of the duodenum and upper jejunum indicates a midgut volvulus.



35.2a,b

Contrast enema gives information only about the position of the cecum, which may occasionally be normally placed even in the presence of a volvulus.

ULTRASONOGRAPHY

Doppler ultrasound of the orientation of the mesenteric vessels can be a useful non-invasive radiological investigation. If the superior mesenteric vein is to the left of the artery, this is diagnostic of malrotation. Other orientations are also known, such as the vein lying anterior to the artery. In some cases of malrotation, however, the orientation between the vessels is normal and this does not exclude malrotation.

The diagnosis of midgut volvulus on ultrasound can be made if a "whirlpool sign" is seen. This is most often seen in cases that present beyond the neonatal period.

Preoperative resuscitation

Patients presenting with acute strangulating obstruction require rapid resuscitation before proceeding to surgery. This comprises rapid IV volume replacement, nasogastric decompression, correction of electrolyte and acid–base imbalance, and administration of broad-spectrum antibiotics. Attempts should be made to correct hypothermia. The period of intensive resuscitation should not extend for more than 1–2 hours before proceeding to surgery, as prolonging the time will expose the intestine to increased ischemia and may result in more extensive bowel necrosis.

OPERATIONS

Surgical correction of the anomaly should always be regarded as an emergency, even in patients presenting non-acutely. Volvulus may supervene at any stage and the operation should be scheduled as early as possible.

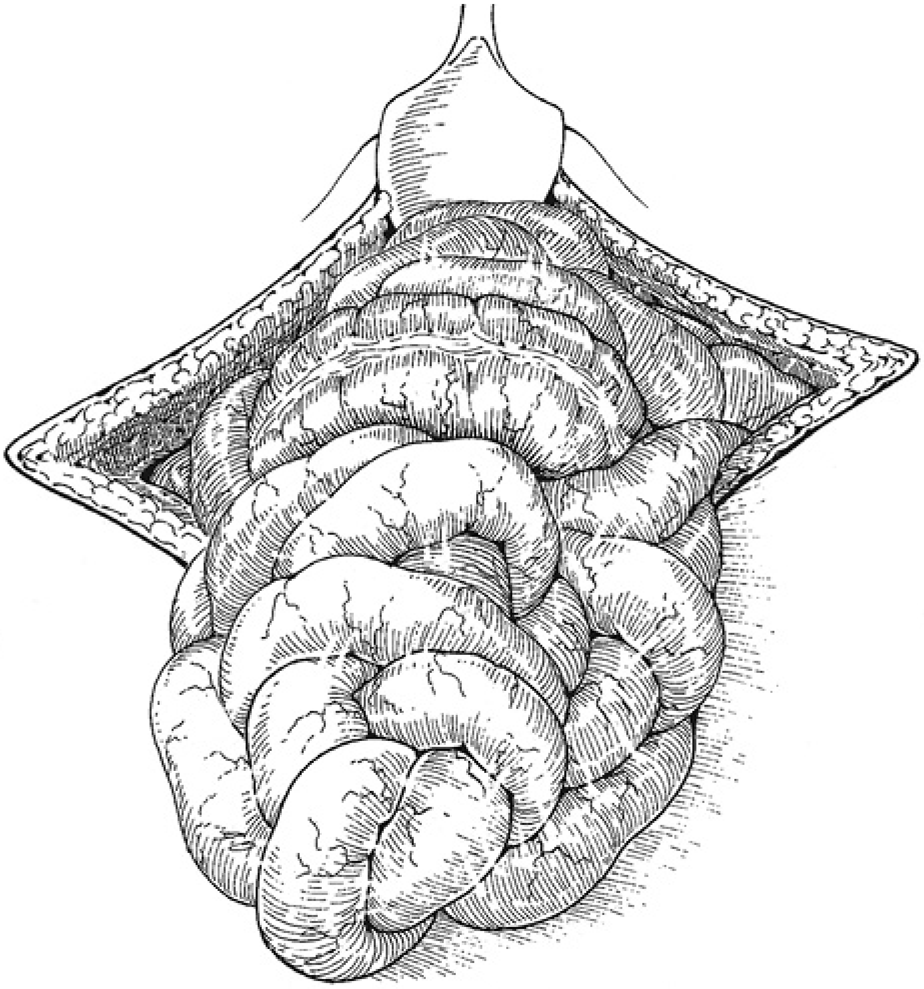
Laparotomy

INCISION

A laparotomy is performed via an upper abdominal, transverse, muscle-cutting incision, extending mainly to the right side. All of the intestines are delivered into the wound for careful examination. A small volume of white to yellowish, free peritoneal fluid (chylous ascites) is usually present in any early intestinal obstruction, but blood-stained fluid is indicative of intestinal necrosis.

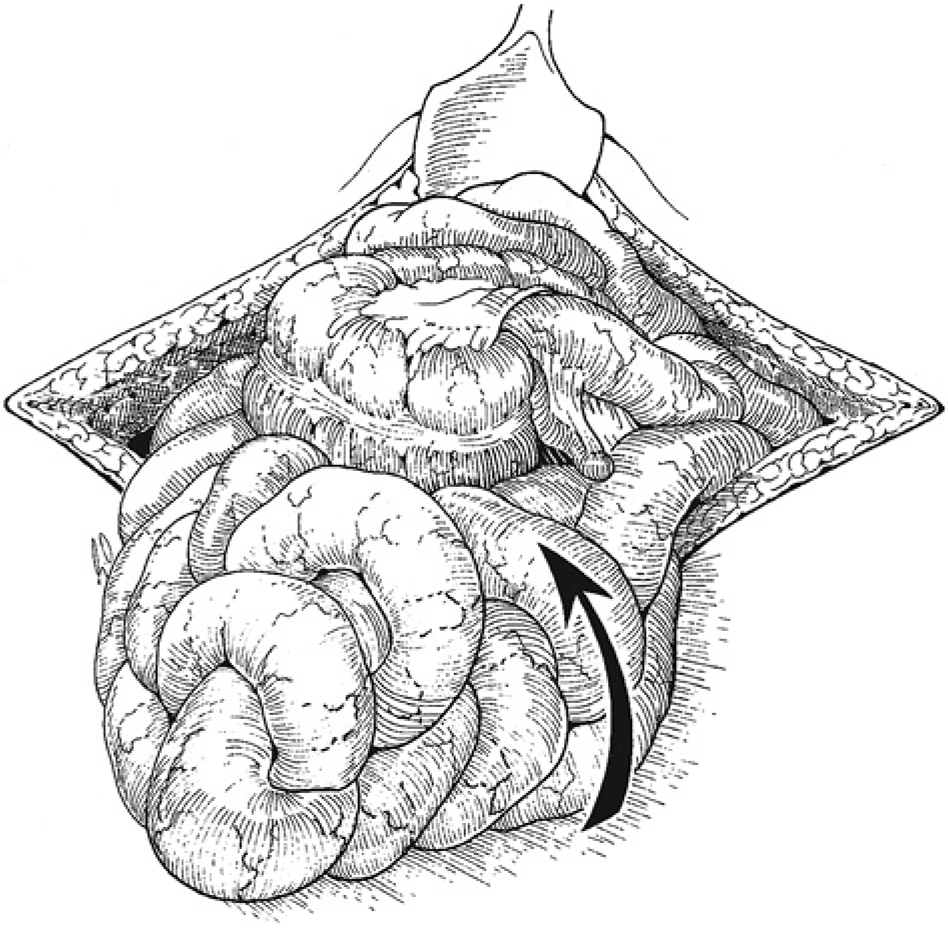
MIDGUT VOLVULUS

The volvulus occurs around the base of the narrow midgut mesentery. The twist usually occurs in a clockwise direction and is untwisted by as many rotations as required (**Figure 35.3**).



35.3

Moderately ischemic bowel, which appears congested and dusky, rapidly resumes a normal pinkish color on reduction of the volvulus. Frankly necrotic bowel is extremely friable and may disintegrate on handling. After untwisting (**Figure 35.4**), bowel of questionable viability should be covered with warm, moist swabs and left undisturbed for about 10 minutes before assessing the extent of ischemic damage. A Ladd's procedure to correct the malrotation is carried out (see next section).



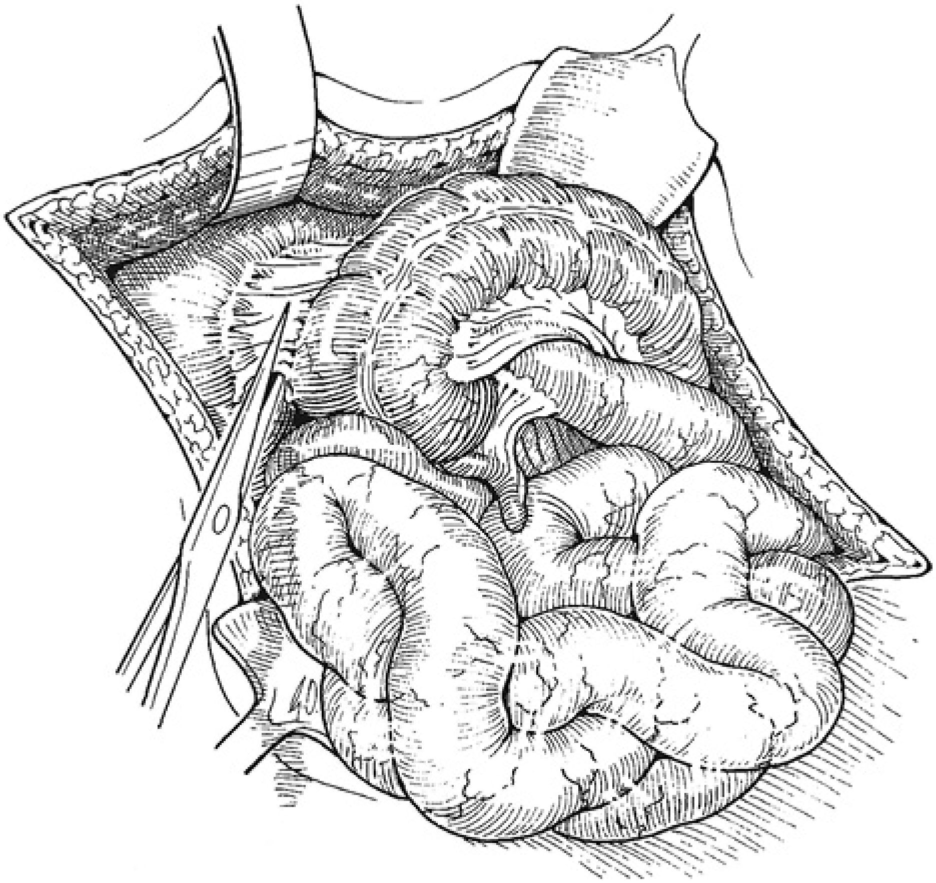
35.4

Frankly necrotic bowel should be resected and the bowel ends either tied off or stomas fashioned with a view to performing a second-look laparotomy in 24–48 hours. Lines of demarcation should be clearly evident at this stage, and an end-to-end anastomosis may be feasible. In the intervening period, the patient is electively ventilated and resuscitative measures continued.

Uncomplicated malrotation (Ladd's operation)

This operation is named after William E. Ladd (1880–1967), the patriarch of American pediatric surgery, who worked in Boston, MA, in the 1920s and 1930s. The aim of the procedure is to restore intestinal anatomy to the non-rotated position with the duodenum and upper jejunum on the right of the abdomen and the cecocolic loop in the LUQ.

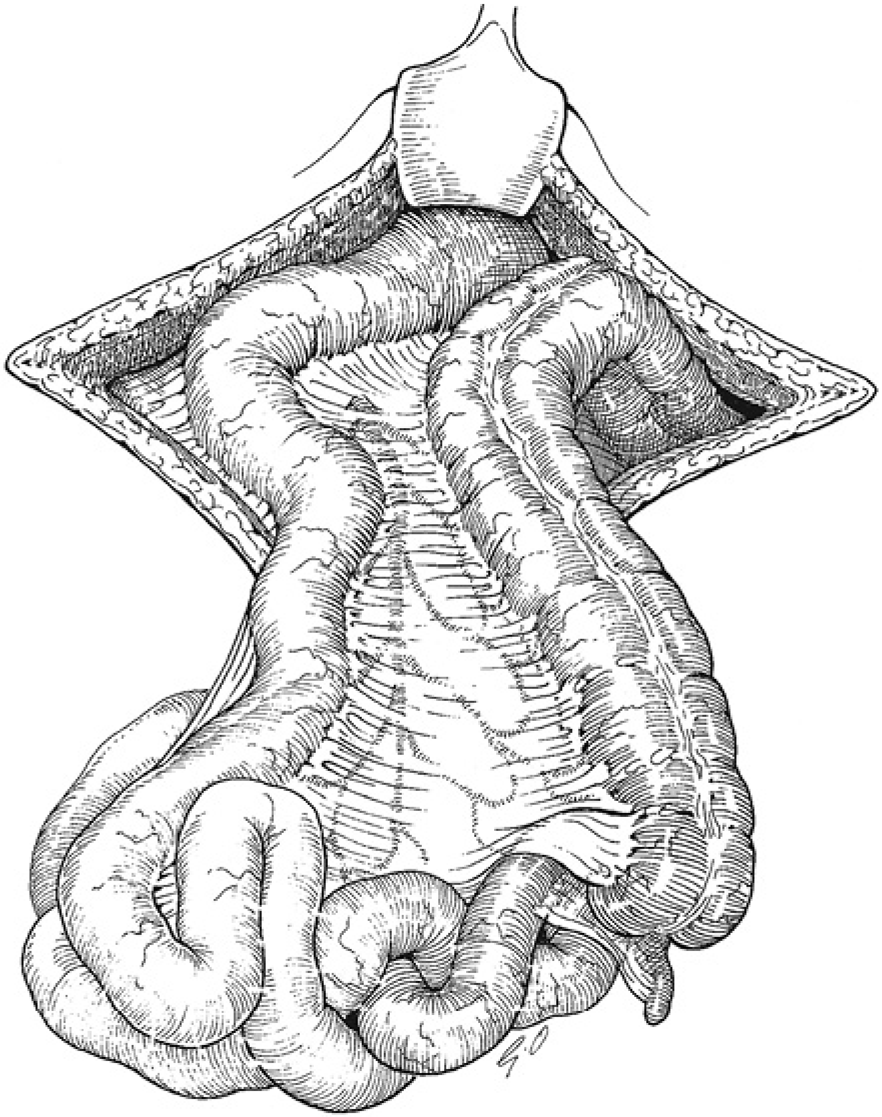
Folds of peritoneum (Ladd's bands) extending from the cecum and ascending colon across the duodenum to the right paracolic gutter and to the liver and gallbladder are carefully divided. This maneuver leaves the cecocolic loop free laterally, but dense adhesions in the base of the mesentery must be divided before the cecum and ascending colon can be fully separated from the duodenojejunal loop. Separation is achieved by opening the serosa of the mesentery between the duodenum and cecum and exposing the anterior surface of the superior mesenteric vessels coursing in the narrow-based mesentery to the midgut (**Figure 35.5**).



35.5

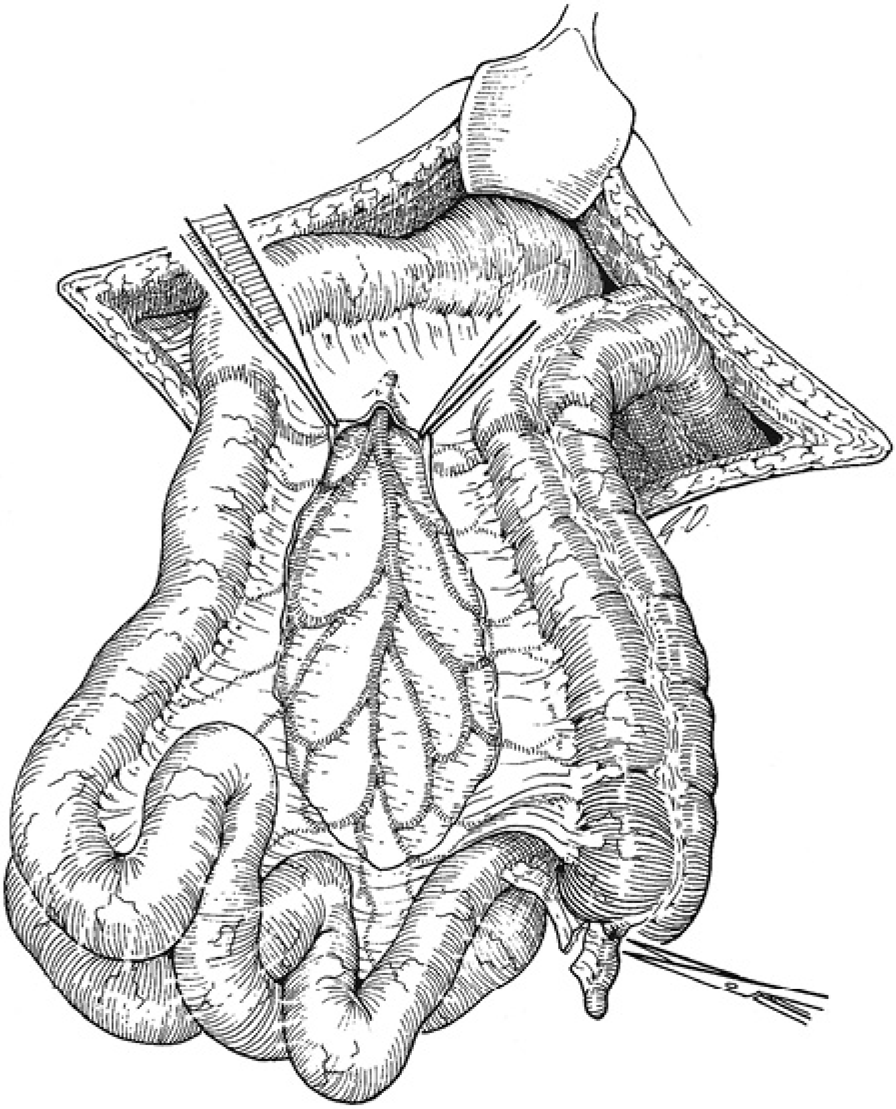
The mesentery in this part is often thickened and edematous, especially if an associated midgut volvulus needs to be untwisted. Care should be taken to avoid trauma to the main vessels, and small branches may need to be ligated before being divided. Large, fleshy lymph and lymphatic channels that have been divided should be sealed by ligation or electrocoagulation to avoid postoperative chylous leakage.

The mesentery is widened peripherally to allow the right colon to be mobilized. Centrally, the dissection is continued into the base of the mesentery until the superior mesenteric artery and vein are freed of any fibrous compression (**Figure 35.6**).



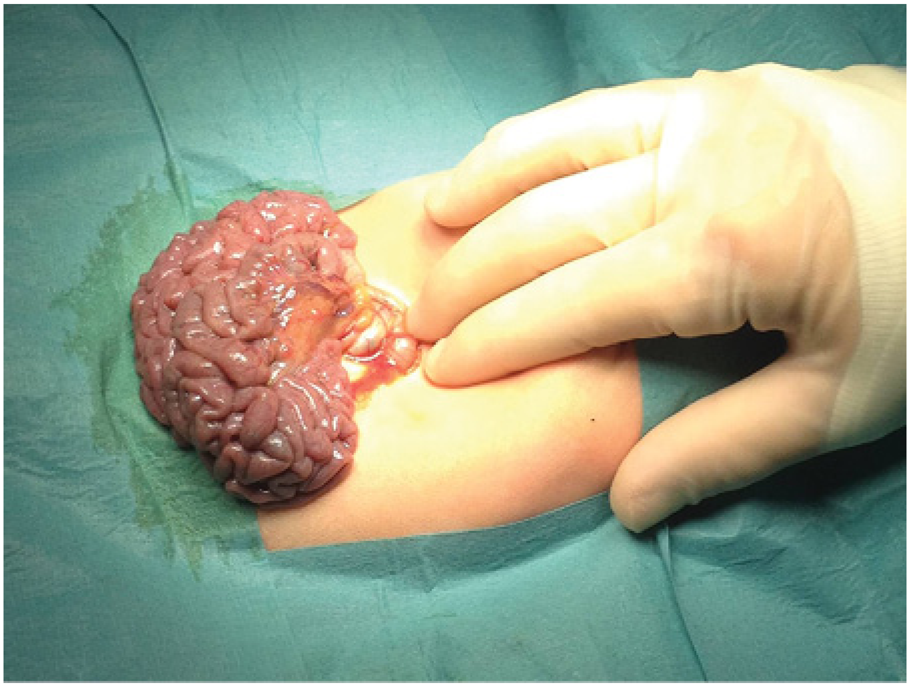
35.6

The duodenum is straightened by dividing the ligament of Treitz, following which the duodenum should be carefully inspected for the presence of any intrinsic obstruction (**Figure 35.7**). If there is any doubt, a balloon catheter should be passed through the duodenum into the proximal jejunum, inflated, and carefully withdrawn into the stomach. Inability to pass the catheter through the duodenum or hold up of the inflated balloon on withdrawal indicates an intrinsic obstruction.



35.7

An appendicectomy may also be performed at this stage as the cecum will be placed in the LUQ of the abdomen and the diagnosis of subsequent appendicitis in later life may be difficult to establish (**Figure 35.8**). Appendicectomy can be performed either as a standard ligation of the appendix base and resection or as an inversion appendicectomy, thus avoiding potential contamination. There are, however, reports of postoperative intussusception arising from inversion appendicectomy.



35.8 Intraoperative photograph taken at laparotomy from the left of the patient showing a midgut volvulus with viable small bowel.

The intestine is replaced in the peritoneal cavity, commencing with the duodenum and proximal jejunum that lie on the right side and ending with the terminal ileum and cecum that are placed in the left hypochondrium. No attempt is made to fix the intestine in this position.

Laparoscopy

Laparoscopy has two potential roles in the management of malrotation:

- It can be used in the treatment of symptomatic malrotation.
- In cases of doubtful malrotation, it can help decide whether or not surgical therapy is required.

TREATMENT OF SYMPTOMATIC MALROTATION

A significant number of series reporting successful laparoscopic correction of malrotation volvulus have now been published, meaning that volvulus is no longer an absolute contraindication to the laparoscopic approach. Laparoscopy in this situation should be limited to hemodynamically stable infants, and parents should be warned about the high risk of conversion to open surgery. Strangulation with necrosis and/or perforation should prompt conversion to an open procedure.

The child is placed in a supine reverse Trendelenburg position on a short operating table. Umbilical access is used for the laparoscope, with working ports on the left and right of the abdomen. The liver may be elevated by means of an additional instrument or a Nathanson retractor.

Once access to the abdomen is established, the diagnosis is confirmed by identifying the position of the duodenojejunal flexure and the ileocecal junction. The volvulus is reduced. The right colon is then reflected to the patient's left to expose the duodenum. At this point, careful division of the attachments between the duodenum and colon proceeds to reveal the superior mesenteric artery. The duodenum is then straightened by division of its right lateral attachments. The small bowel can then be brought to the right of the abdomen by gentle manipulation into the right upper quadrant, progressing from the duodenal end to the ileocecal junction. Peritoneal attachments restricting this are divided as they are encountered. This rewards the operator with a view of the small bowel mesentery with the small bowel on the right of the image and the colon to the left. Final division of the attachments between the small bowel and colon establishes a non-rotated position. Appendectomy can then proceed if desired.

DOUBTFUL MALROTATION

The diagnosis of malrotation may be difficult to make. The position of the duodenojejunal flexure on upper gastrointestinal study may be just on the left pedicle and not clearly to right or left. An inverted relationship between the superior mesenteric artery and vein may be present on ultrasound. The critical question is not whether there is a disorder of rotation, but whether the mesenteric stalk is narrow and not fixed to the posterior abdominal wall. In other words, is there a high likelihood of volvulus? Laparoscopy is an ideal tool to look at the broadness and the fixation of the mesenteric stalk.

More often than not, the diagnosis of "dangerous malrotation" is excluded.

OUTCOME

Recovery is generally prompt and uncomplicated. Infants who have suffered extensive bowel loss following midgut volvulus may clearly experience problems of short bowel syndrome with a prolonged requirement for parenteral nutrition.

It is felt that performing an open Ladd's procedure may help to promote adhesions that effectively prevent recurrent volvulus, but adhesive intestinal obstruction is relatively common, with short-term rates as high as 10%, and higher incidences reported with longer follow-up. Conflicting evidence on the rate of recurrent volvulus following laparoscopic Ladd's procedure exists in the literature with some series showing higher rates and others lower when compared to the open counterpart.

Our recently reported experience of 65 cases (**Table 35.1**) shows a 6% rate of redo surgery, with all of these cases within the early part of the experience. Some surgeons perform a cecopexy at a laparoscopic procedure to promote adhesions to the cecum with the intention of preventing recurrent volvulus, although this is not routinely practiced. The technically challenging nature of the laparosopic approach is associated with a high conversion rate of at least 15%, which is higher both in younger infants and in those with volvulus.

Table 35.1 **Great Ormond Street experience with laparoscopic Ladd's procedure (2007–2017) (n = 65)**

	Volvulus (n = 16)	Malrotation without volvulus (n = 29)	Asymptomatic malrotation (n = 20)	P value
Birth weight (kg)	3.3 (0.7–3.8)*	3.1 (1.52–4.06)	3.0 (1.0–3.6)	0.12
Age at surgery (days)	43 (1–2373)	232 (3–5565)	228 (1–5780)	0.19
Weight at surgery (kg)	3.7 (2–20.3)	8.3 (2.55–10.8)	5.6 (2–29.2)	0.08
Associated anomalies	8 (50%)	22 (76%)	20 (100%)	
Conversion to open	6 (38%)	2 (7%)	2 (10%)	
Time to full feeds (days)	7 (3–27)	4 (1–23)	2.5 (1–8)	0.0002

Source: Modified from Arnaud et al. (2019).

* Median (range).

As more and more minimal invasive surgical techniques are introduced into pediatric and neonatal surgery, there is ongoing debate about management for intestinal rotational abnormalities associated with other defects, such as congenital diaphragmatic hernias and abdominal wall defects. In the context of such abnormalities, it is recommended that an upper gastrointestinal contrast study be undertaken, and, if malrotation is diagnosed, that a laparoscopic examination of the intestine be performed and Ladd's procedure undertaken if required.

FURTHER READING

Arnaud AP, Suply E, Eaton S, et al. Laparoscopic Ladd's procedure in infants and children is still a controversial approach. *J Pediatr Surg* 2019; 54: 1843–7.

El-Gohary Y, Alagtal M, Gillick J. Long-term complications following operative intervention for intestinal malrotation: A 10-year review. *Pediatr Surg Int* 2010; 26: 203–6.

Fraser JD, Aguayo P, Sharp SW. The role of laparoscopy in the management of malrotation. *J Surg Res* 2009; 156: 80–82.

Lampl B, Levin TL, Berdon WE, Cowles RA. Malrotation and midgut volvulus: A historical review and current controversies in diagnosis and management. *Pediatr Radiol* 2009; 39: 359–66.

Skandalakis JE, Gray SW. *Embryology for Surgeons*, 2nd ed. Baltimore, MD: Williams & Wilkins, 1994.

Stanfill AB, Pearl RH, Kalvakuri K, et al. Laparoscopic Ladd's procedure: Treatment of choice for midgut malrotation in infants and children. *J Laparoendosc Adv Surg Tech* 2010; 20: 369–72.

36

Congenital atresia and stenosis of the intestine

SHARON G. COX, ALASTAIR J.W. MILLAR, AND ALP NUMANOGLU

HISTORY

Jejunoileal atresia, defined as a congenital defect of intestinal luminal continuity, is a common cause of obstruction in the newborn.

In 1911, Fockens of Rotterdam reported the first successfully treated case of small intestinal atresia. Mortality remained high, though, and in a comprehensive review of the world literature up to 1950, Evans could find reports of only 39 successfully treated cases of jejunoileal atresia.

In 1952, Janie Louw from Cape Town suggested that most jejunoileal atresias were probably due to a vascular accident rather than being the result of inadequate recanalization, as had previously been thought. At his instigation, Christian Barnard perfected an experimental model in pregnant mongrel bitches and reproduced all types of atresia found in humans. This not only confirmed Louw's hypothesis, but also provided the opportunity to improve the technical aspects of corrective surgery, which involved resection of the dilated proximal blind-ending bowel and primary end-to-end anastomosis, resulting in improved outcomes.

The management of jejunoileal atresia has been one of the great success stories of pediatric surgery of the 20th century. Significant changes to the surgical technique made during the 1950s resulted in survival rates increasing from 10% to over 80%.

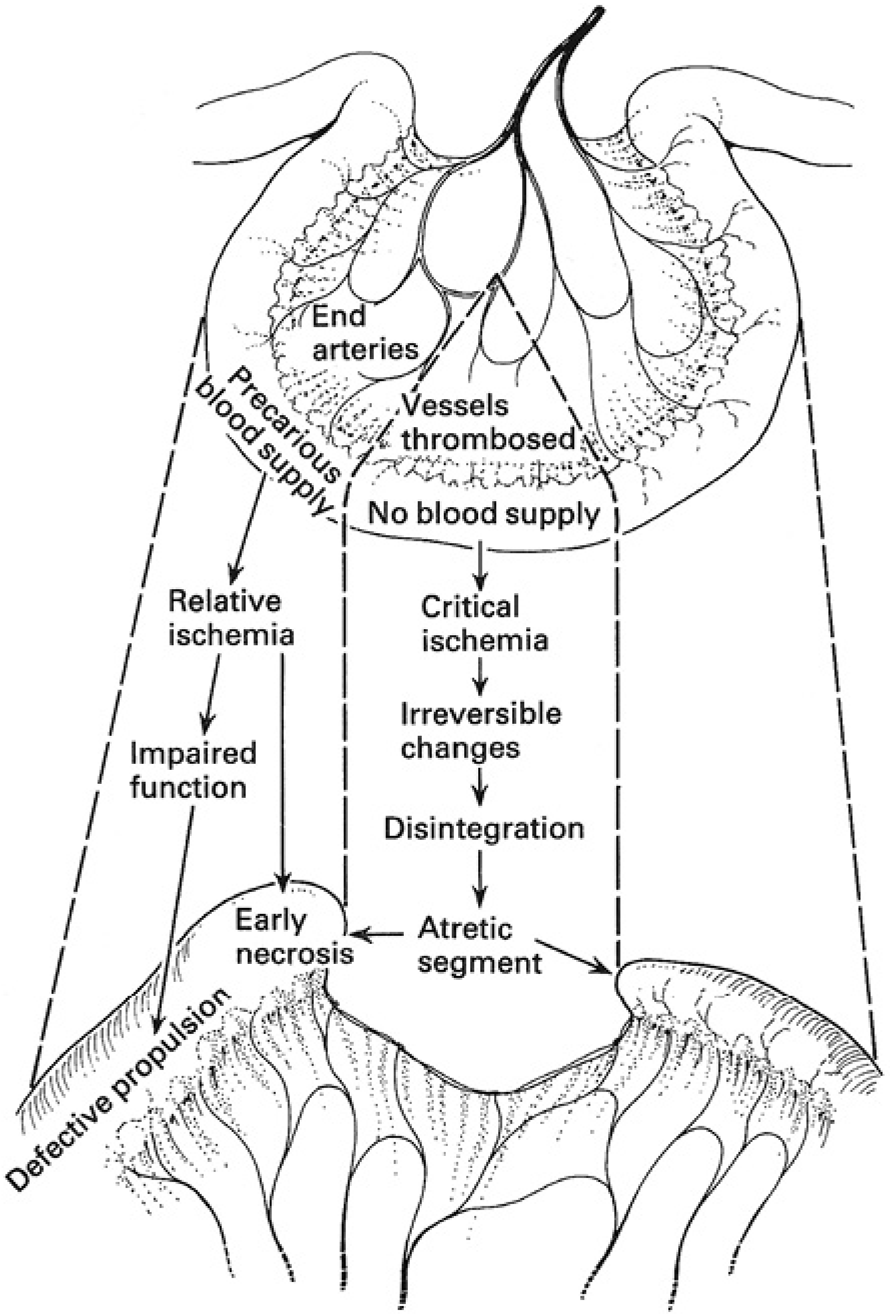
PRINCIPLES AND JUSTIFICATION

Incidence

The incidence of jejunoileal atresia varies from 1 in 1500 to 1 in 3000 live births.

Pathogenesis

Although several mechanisms have been postulated, the most feasible theory is that of a localized intrauterine vascular incident resulting in ischemic necrosis, liquefaction, and absorption of the affected devascularized segment(s). The ischemic hypothesis is further supported by additional evidence of incarceration or snaring of bowel in an exomphalos or gastroschisis, fetal intussusception, midgut volvulus, transmesenteric hernias, and thromboembolic occlusions. The anomaly is usually not genetically determined. The ischemic insult causes morphologic and functional abnormalities of the remaining proximal and distal bowel. The blind-ending proximal bowel becomes dilated and hypertrophied, resulting in functional abnormalities that include ineffective peristalsis. The distal bowel is unused and collapsed, but with potential normal length and function (**Figure 36.1**).


End
arteries
Precarious
blood supply
Vessels
thrombosed
No blood supply
Relative
ischemia
Critical
ischemia
Irreversible
changes
Impaired
function
Disintegration
Early
necrosis
Atretic
segment
Defective propulsion

36.1

The etiology of some familial multiple atresia syndromes may be due to genetic mutations (*TTC7A*) and are associated with immune deficiency syndromes (MIA-SCID). These are typically multiple Type I atresias within the small intestine with the most proximal being a pyloric atresia. It is usually a fatal condition without small intestinal and bone marrow transplantation.

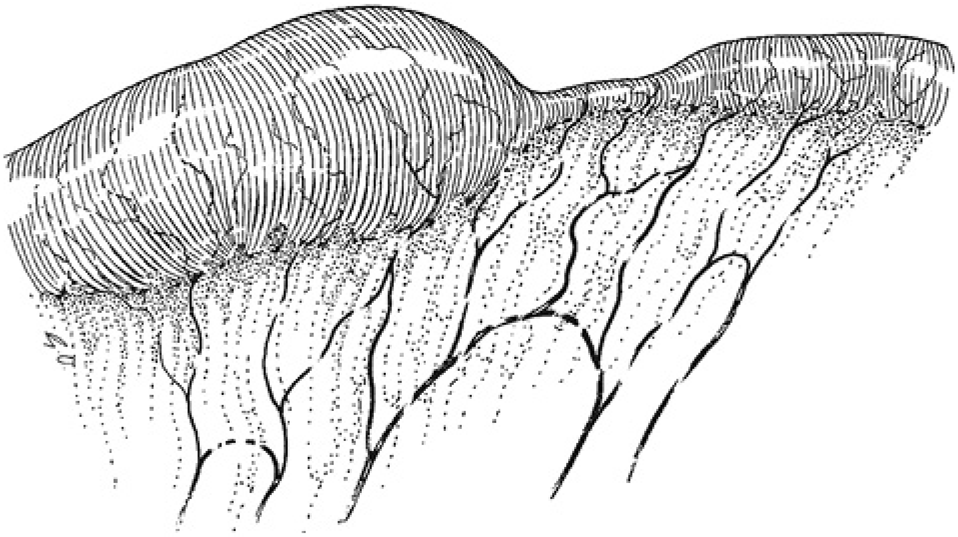
Classification

The classification of intestinal atresia is well established and covers a spectrum from a mild stenosis to multiple atresias resulting in significant short bowel.

The most proximal atresia or stenosis determines whether it is classified as jejunal or ileal. The current classification is a derivative of the original Bland-Sutton classification (Types I, II, and III), named after English surgeon Sir John Bland-Sutton (1855–1936), with the addition of Type IV to designate multiple atresias, as suggested by Jay Grosfeld, American pediatric surgeon (1935–2016).

STENOSIS

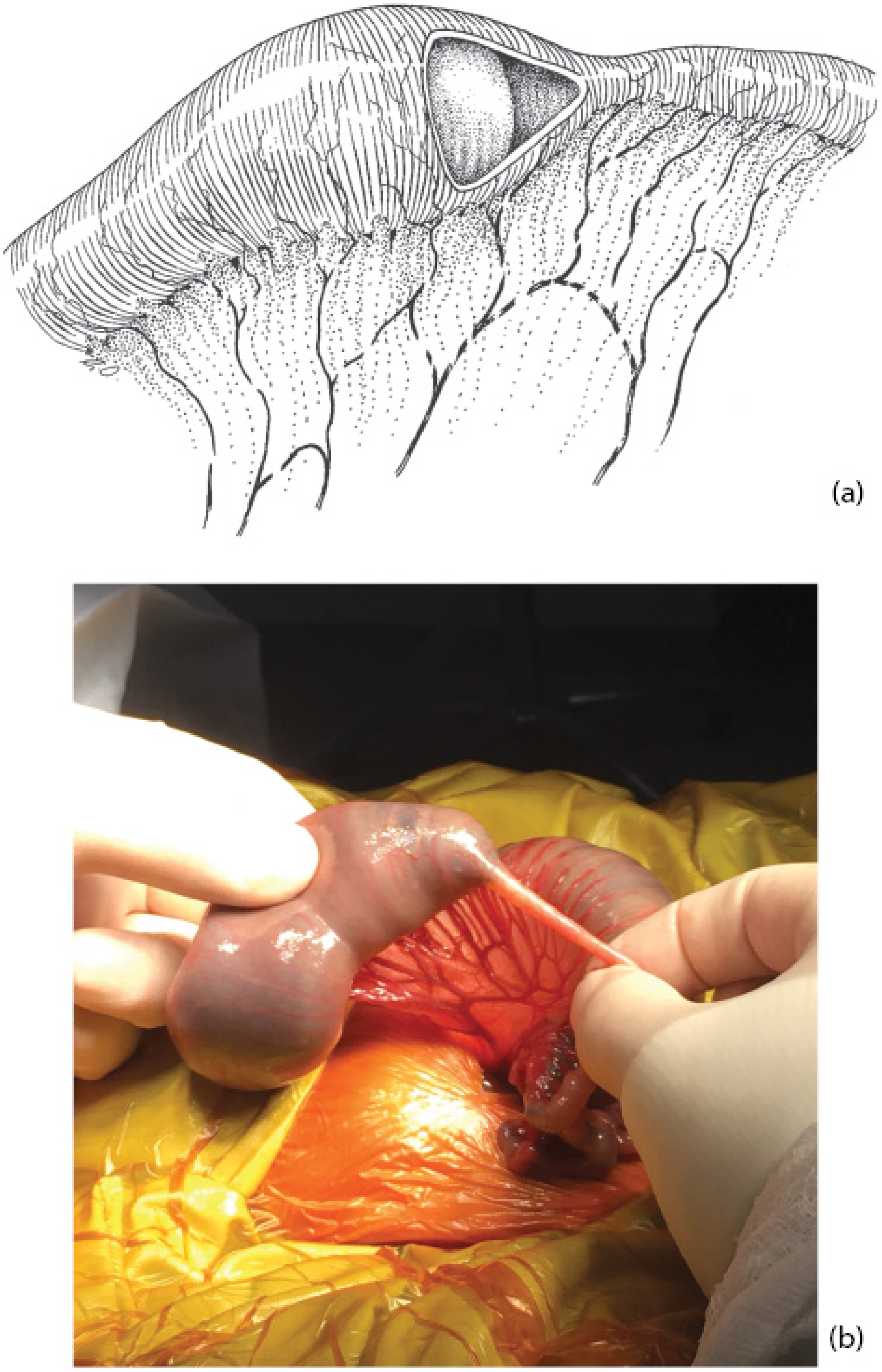
The proximal dilated and distal collapsed segments of intestine are in continuity with an intact mesentery, but at the junction there is a short, narrow, somewhat rigid segment with a minute lumen which may mimic atresia Type I (**Figure 36.2**). The small intestine is of normal length.



36.2

ATRESIA TYPE I (MEMBRANE)

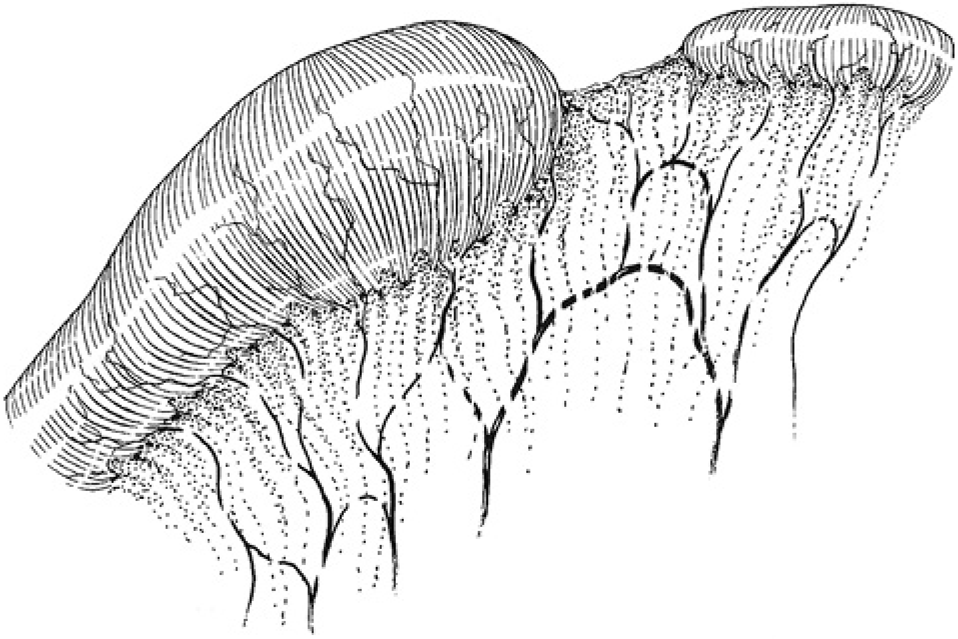
The dilated proximal and collapsed distal segments of intestine are in continuity and the mesentery is intact. The pressure in the proximal intestine tends to expand the membrane into the distal intestinal lumen, so that the transition from distended to collapsed intestine is conical in appearance, giving the “windsock” effect. The distal intestine is completely collapsed but the bowel immediately distal to the obstruction may be dilated by the windsock (**Figure 36.3a and b**). The small intestine is of normal length.



36.3a,b

ATRESIA TYPE II (BLIND ENDS JOINED BY A FIBROUS CORD)

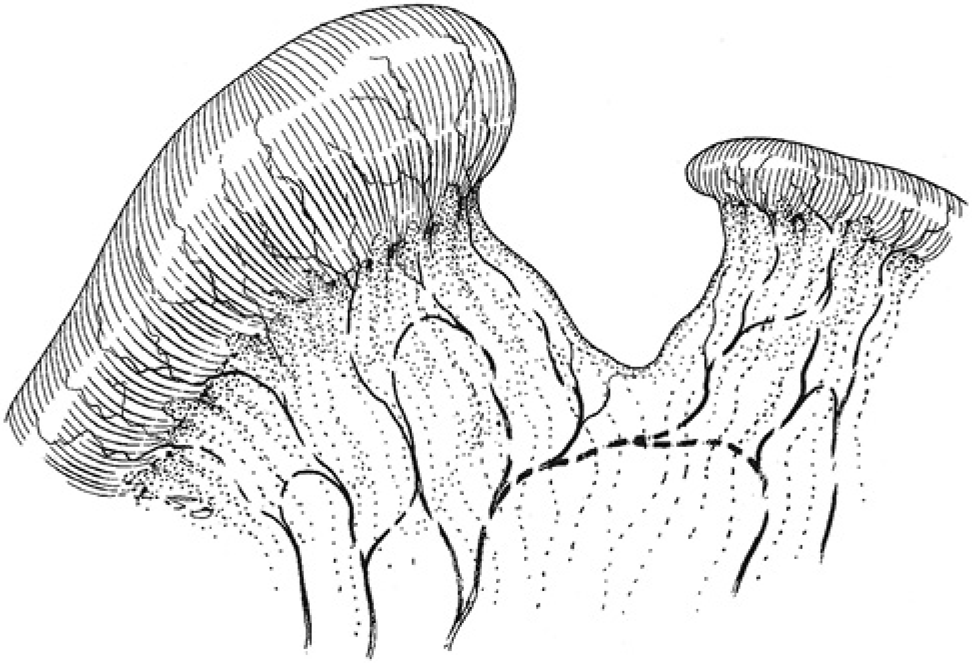
The proximal intestine terminates in a bulbous blind end that is grossly distended and hypertrophied for several centimeters, but more proximally assumes a normal appearance. This blind end is often hypoperistaltic. The distal completely collapsed intestine commences as a blind end that is occasionally bulbous, owing to the remains of a fetal intussusception. The two blind ends are joined by a thin, fibrous band, with the corresponding intestinal mesentery intact (**Figure 36.4**). The small intestinal length is usually normal.



36.4

ATRESIA TYPE IIIA (DISCONNECTED BLIND ENDS)

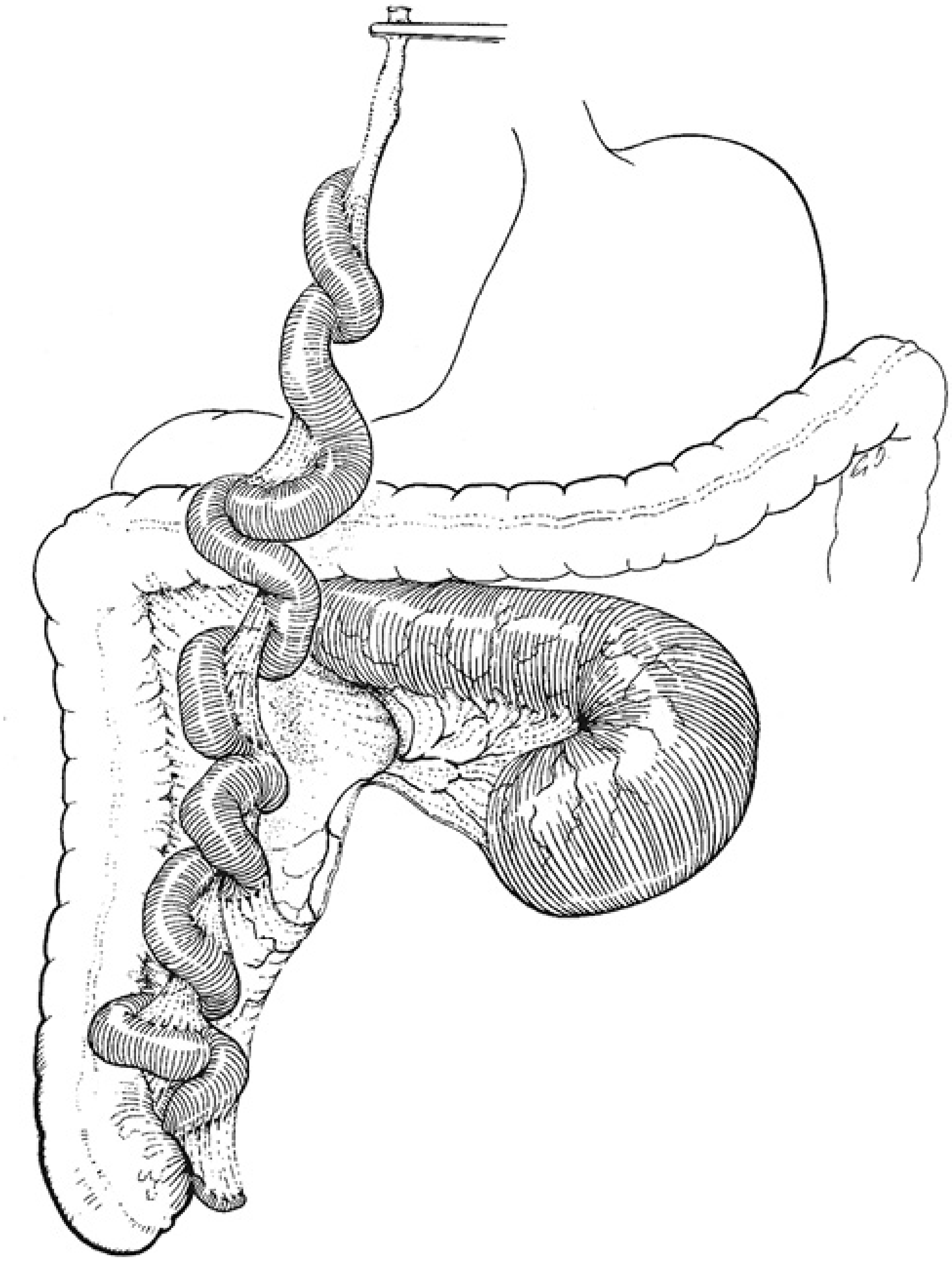
The appearance is similar to that in Type II, but the blind ends are completely separate (**Figure 36.5**). There is always a mesenteric defect of varying size, and the proximal intestine may, as a secondary event, undergo torsion or become overdistended, resulting in ischemia, necrosis, and perforation. The total length of intestine is reduced to a varying extent.



36.5

ATRESIA TYPE IIIB ("APPLE PEEL," "CHRISTMAS TREE" ATRESIA)

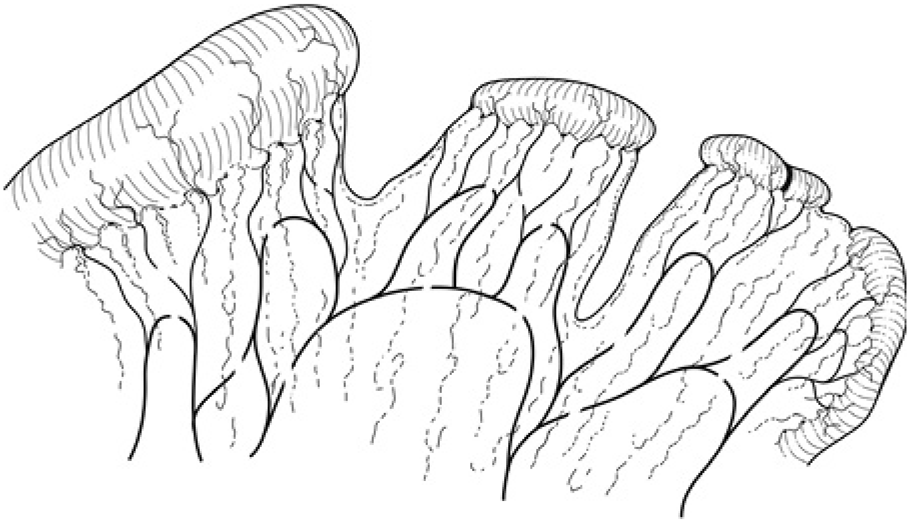
As in Type IIIa, the blind ends are unconnected and the mesenteric defect is large. The atresia is usually localized in the proximal jejunum near the ligament of Treitz, with absence of the superior mesenteric artery beyond the origin of the middle colic branch and absence of the dorsal mesentery. The distal intestine assumes a helical configuration around an attenuated, single artery of blood supply arising from the ileocolic or right colic arcade (**Figure 36.6**). Occasionally, further atresias of Type I or II are found in the distal intestine, usually close to the blind end. Vascularity of the distal intestine may be impaired. There is always a significant reduction in intestinal length.



36.6

ATRESIA TYPE IV (MULTIPLE ATRESIA)

Multiple atresias can be combinations of Types I–III and often present morphologically as a string of sausages (**Figure 36.7**).



36.7

A much more severe condition, usually termed (hereditary or familial) multiple intestinal atresia, is characterized by a multiplicity of Type I (membranes) throughout the small bowel and also sometimes associated with a pyloric atresia. This is due to a genetic mutation (tetratricopeptide repeat domain of *7A* gene). Affected infants invariably have a combined immunodeficiency and are only ever effectively treated by bone marrow and small bowel transplant.

PREOPERATIVE ASSESSMENT AND MANAGEMENT

Clinical presentation

Many cases of intestinal atresia are now being diagnosed prenatally by ultrasonographic and magnetic resonance imaging (MRI) of the fetus, showing dilated intestinal loops and increased echogenicity suggesting obstruction. The higher the obstruction, the more likely there is to be polyhydramnios. Postnatally, atresia or severe stenosis of the small intestine presents as neonatal intestinal obstruction with persistent bile vomiting from the first or second day of life, and varying degrees of abdominal distension. Erythema of the abdominal wall, tenderness, distension, and rigidity may signify bowel ischemia or peritonitis.

Radiology

Abdominal radiographs done 6–8 hours after birth will show distended, air-filled small intestinal loops and a gasless distal abdomen. If incomplete with evidence of distal intestinal gas, an upper gastrointestinal contrast study may be indicated to show the site and nature of the obstruction. If the original plain radiograph suggests a low obstruction, then a contrast enema is suggested to exclude colonic atresia or other causes, such as Hirschsprung disease (especially if total colonic) or meconium ileus. In the latter, there may be features of a right lower quadrant speckling caused by gas admixing with luminal content (soap-bubble or Neuhauser's sign).

Patient preparation

- Nasogastric tube to prevent aspiration and decompress stomach and intestine
- IV fluid management appropriate to age, weight, and hydration state of infant
- Correction of coagulation defects (vitamin K)
- Antibiotics, typically penicillin and a further broad-spectrum antibiotic (e.g. gentamicin, cephalosporin)

OPERATION

The operative procedure depends on the type of atresia. After the initial exploration, it is important to determine the site of atresia as well as presence or absence of the associated atresias. Resection of the dilated proximal segment, meticulous anastomosis keeping in mind the necessity of adequate bowel length during surgery forms the mainstay of surgical management. Type IIIB and Type IV atresias are particularly challenging in this regard.

Principles of surgery

- The whole length of the small intestine should be inspected carefully to determine the site and type of atresia and the most likely pathogenesis.
- Patency of the distal small and large bowel must be confirmed by injection of saline into the bowel distal to the most proximal obstruction. Contrast enema is useful to confirm colonic lumen patency.
- The preferred procedure is resection of a varying length of the bulbous blind end of proximal dilated bowel and a short segment of the collapsed distal bowel with primary, single-layer, end-to-end (end-to-back) anastomosis using interrupted 5/0 or 6/0 polydioxanone (PDS□) monofilament sutures with or without tapering or plication.
- Residual bowel length must be documented.
- Ischemic or twisted bowel must be untwisted, and anastomosis may be delayed for 24 hours to assess viability.
- Every effort should be made to preserve bowel length in the presence of foreshortened bowel in Type III or Type IV atresia.
- Proximal or distal stomas are rarely indicated.
- Bowel must be returned into the abdominal cavity, avoiding twisting or kinking of the anastomosis.
- Additional steps may include derotation of the proximal jejunum and duodenum in high intestinal atresia, with back resection of the dilated bowel followed by tapering or inversion plication of the megaduodenum in the proximal atresia.
- Bowel-lengthening procedures should preferably not be performed at the initial operation.

Incision

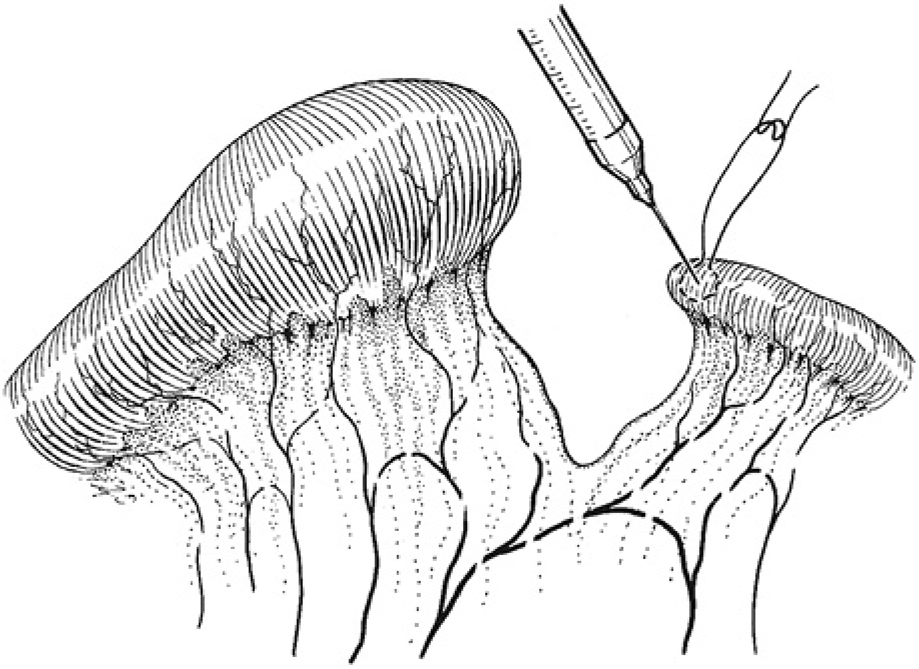
Traditionally, adequate exposure is obtained through a supraumbilical, transverse incision transecting the rectus muscles 2 cm above the umbilicus. Increasingly, minimally invasive techniques are used with small circumumbilical incisions and extracorporeal anastomosis.

Exploration

In uncomplicated cases, the intestine can be delivered through the wound by gentle pressure applied to the abdominal wall. If free gas escapes on opening the peritoneum, or if there is contamination of the peritoneal cavity, the perforation should be identified immediately and closed before further exploration. The intestine proximal to the obstruction is distended, whereas the intestine distal to the obstruction is collapsed, wormlike, and may contain intraluminal contents. The intestine is exteriorized to determine the site and type of obstruction and to exclude other areas of atresia or stenosis, as well as associated lesions, such as incomplete intestinal rotation and meconium ileus.

Detection of other atresias

The distal intestine is carefully examined after the type and location of the atresia have been determined. If a volvulus is present, the bowel should be untwisted and examined carefully for other atretic or stenotic segments, which can occur in up to 20% of cases. Intraluminal membranes are best detected and localized by inserting a catheter or needle into the lumen of the collapsed intestine injecting saline and following the fluid down to the cecum (**Figure 36.8**). Colonic atresia is excluded by a similar procedure through the cecum or by a previously performed contrast enema.

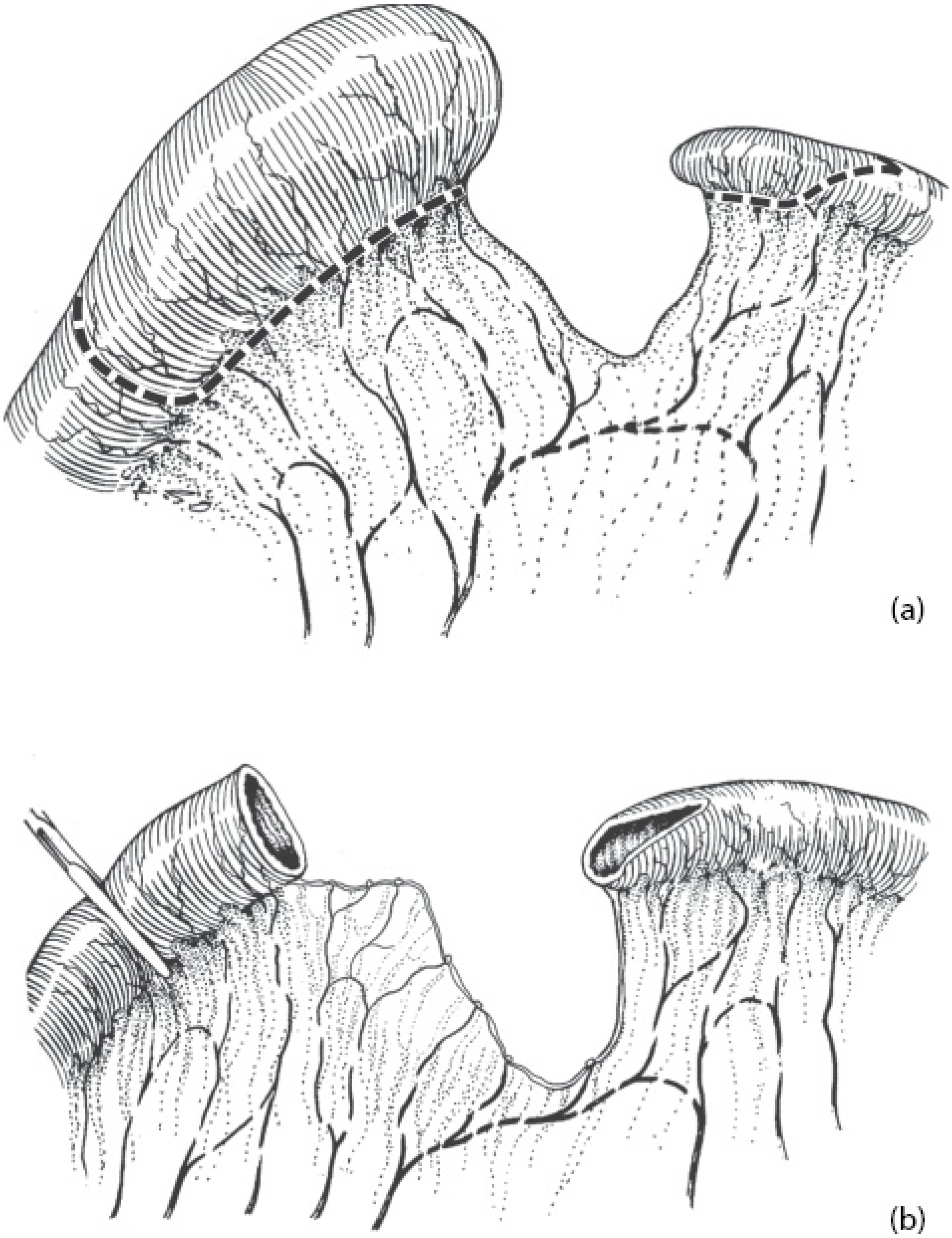


36.8

The total length of unstretched small intestine should be measured accurately along the antimesenteric border. The overall intestinal length at birth in the term infant is about 300 cm (of which 80% is small intestine) but is considerably less in the very preterm infant at around 110–170 cm.

Resection

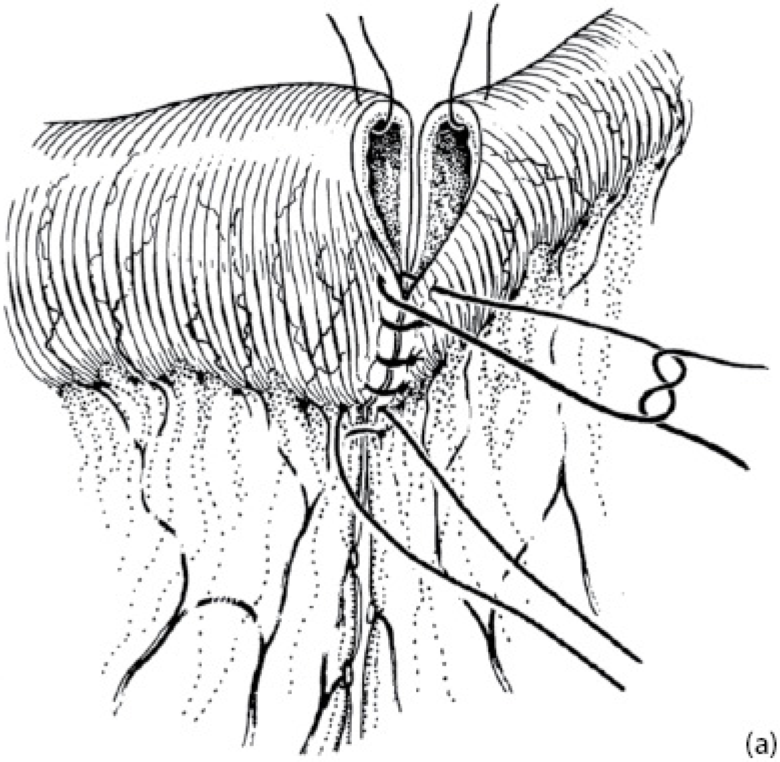
The atretic area and adjacent distended and collapsed loops of intestine are isolated. After milking the intestinal contents into the proximal bulbous end, or in high jejunal atresia into the stomach, from where it is aspirated, an atraumatic bowel clamp is applied across the bowel a few centimeters proximal to the site selected for transection. To ensure adequate postoperative function, the proximal distended and hypertrophied intestine should be liberally resected. Usually, 10–15 cm of bowel is resected even if it appears viable (**Figure 36.9a**). The mesentery adjoining the portion to be resected should be preserved as much as possible by doing the division as close as possible to the bowel wall. It is either divided between clamps and ligated or divided using bipolar diathermy. The proximal intestine is transected at right angles. The blood supply at this level should be excellent. Approximately 2–3 cm of the distal intestine is then resected using a slightly oblique line of transection to create a "fish mouth" that renders the opening about equal in size to that of the proximal intestine (**Figure 36.9b**). Alternatively, an extramucosal end-to-end anastomosis is possible with diameter discrepancy of up to 8 : 1. A culture swab is taken from the proximal gastrointestinal tract, which may have become colonized with bacteria.



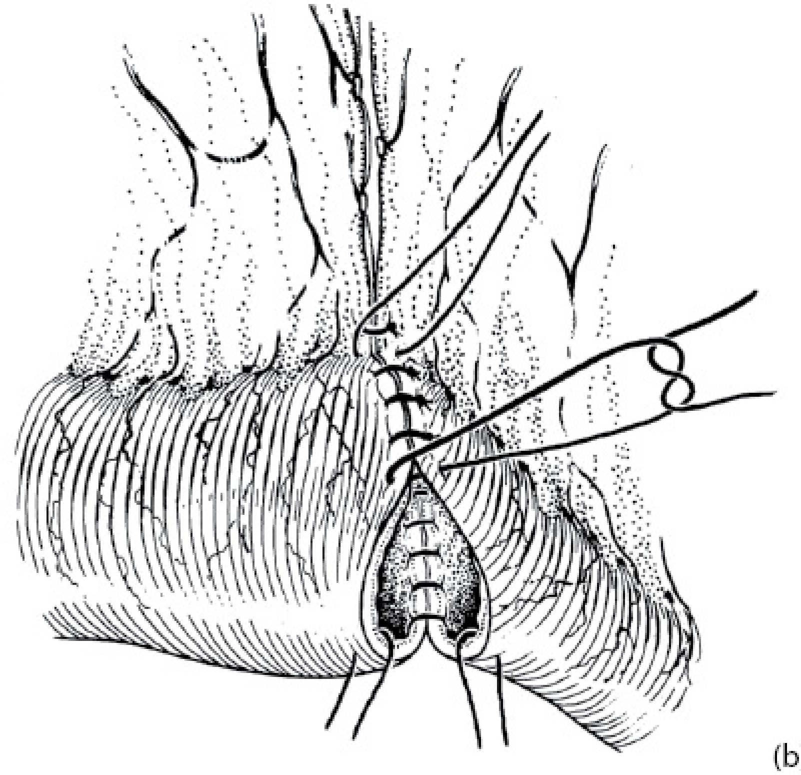
36.9a,b

Anastomosis

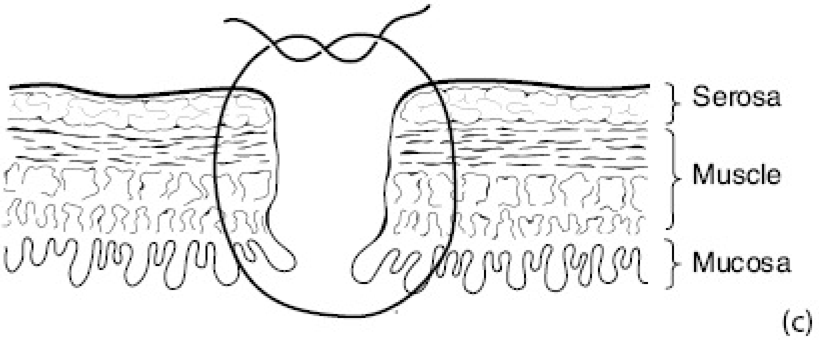
The “anterior” bowel edges are joined by interrupted full thickness sutures, starting from the mesenteric side, and tied on the serosal surface (**Figure 36.10a–c**). Once the anterior layer has been completed, the bowel is rotated 180 degrees to expose the back wall. If there is size discrepancy between the transected proximal and distal ends, a short cut back along the antimesenteric border can be performed on the distal bowel.



(a)



(b)

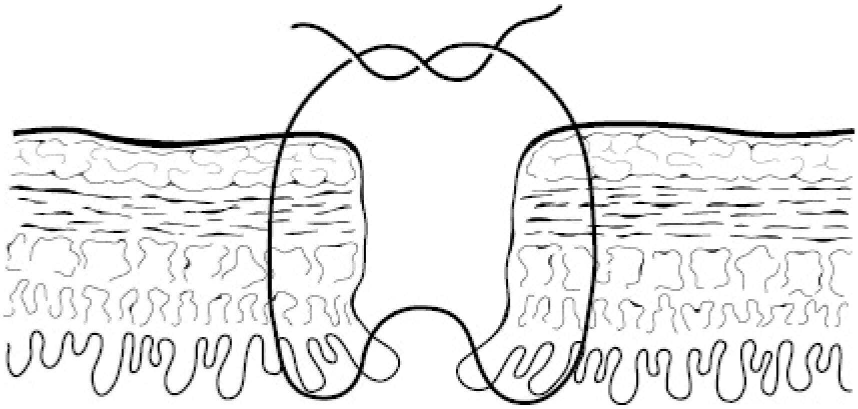


36.10a–c

The same technique can be used for stenoses and intraluminal membranes. Procedures such as simple enteroplasties, excision of membranes, and bypassing techniques are not recommended because they fail to remove the abnormal segment of intestine. Side-to-side anastomosis is avoided because of the increased risk of creating blind loops.

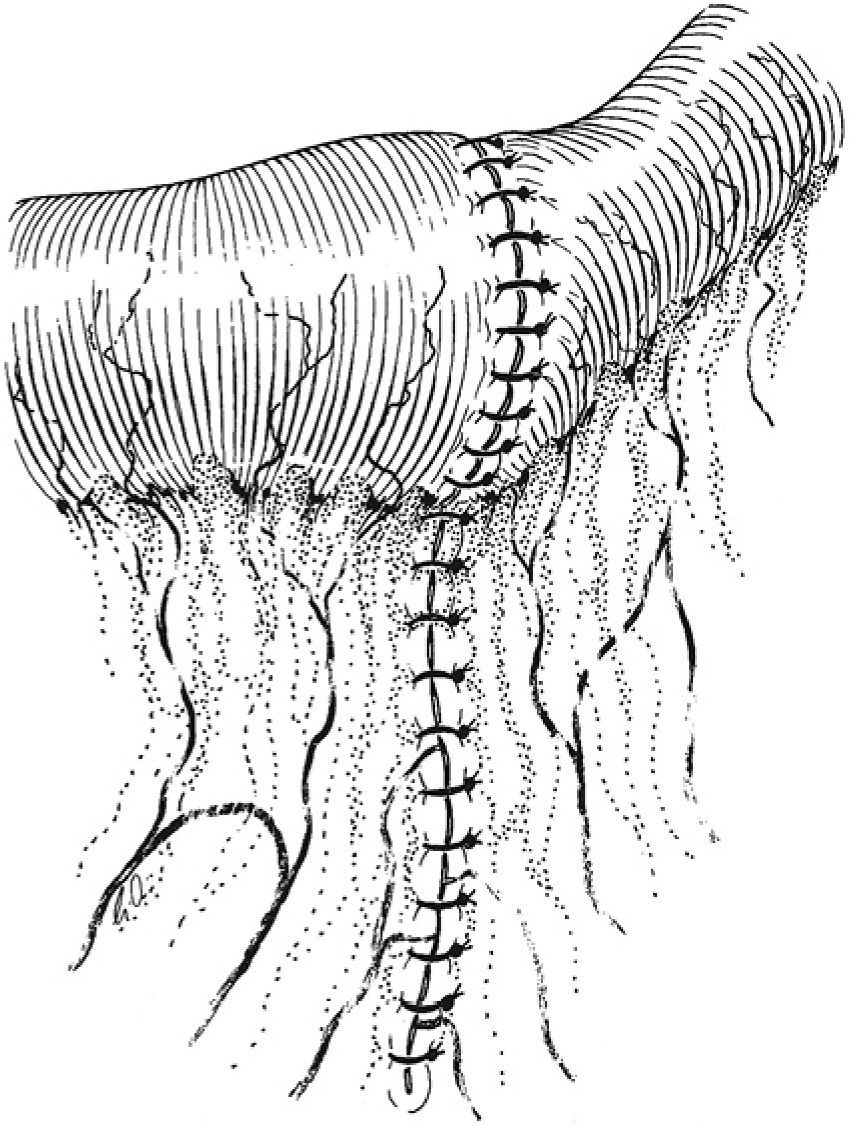
Completion of anastomosis and closure of mesenteric gap

Gambee interrupted inverting or extramucosal stitches may also be used instead of through-and-through stitches (**Figure 36.11**). This facilitates a more end-to-end anastomosis with size discrepancies of up to 8 : 1 accommodated. Alternatively, the posterior bowel edges are united with interrupted through-and-through or inverting Gambee sutures, with the knots tied on the mucosal surface. The anterior bowel edges are then joined in a similar fashion, with the knots being tied on the serosal surface.



36.11

The suture lines are inspected and additional stitches are placed if required to ensure a “watertight” anastomosis. The defect in the mesentery is repaired by approximating (and overlapping if necessary) the divided edges with interrupted sutures, taking great care not to kink the anastomosis or to compromise the blood supply (**Figure 36.12**). Thereafter the intestines, well moistened with warm saline, are returned to the peritoneal cavity.



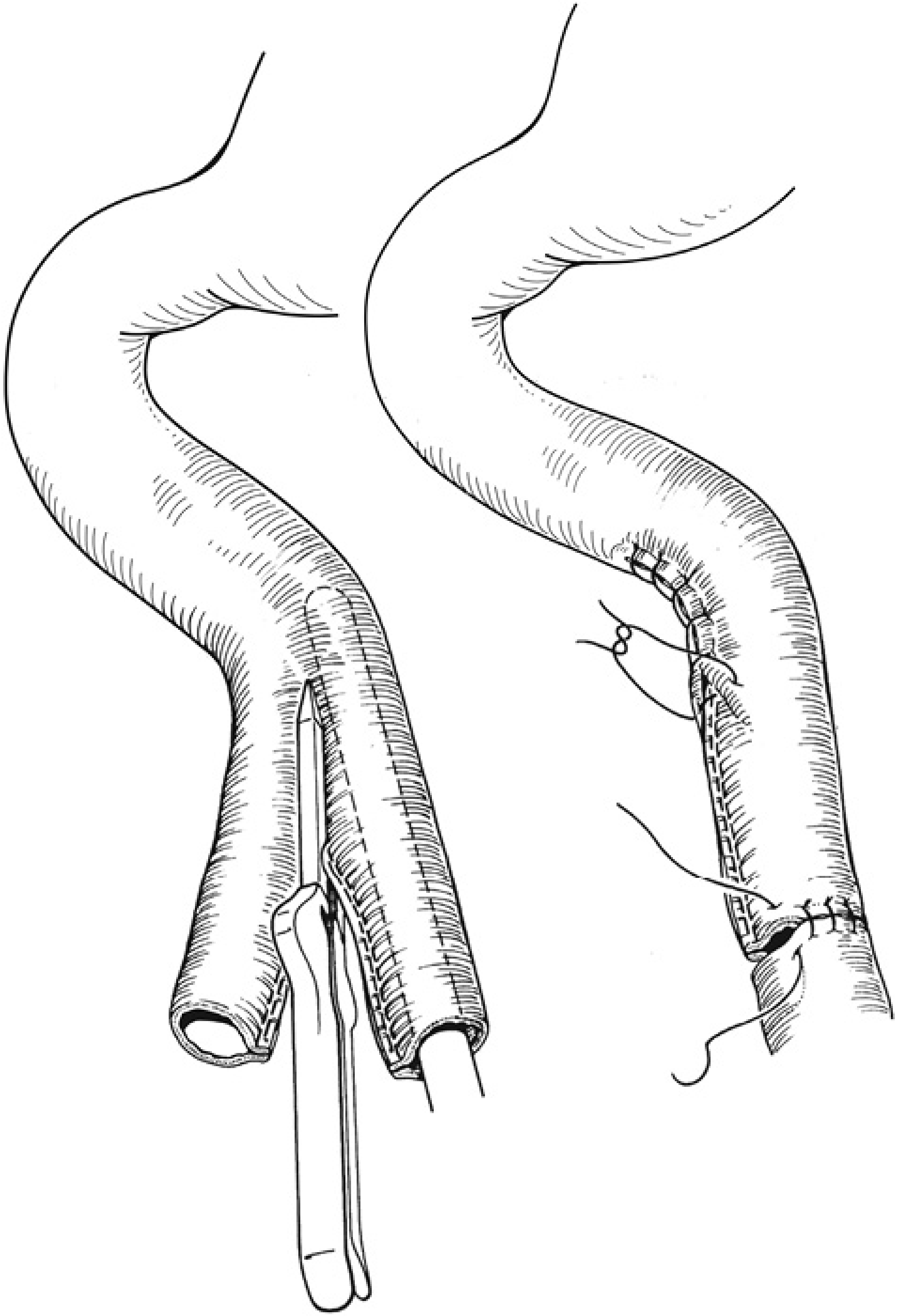
36.12

BOWEL-SAVING PROCEDURES

Tapering duodenojejunoplasty or enteroplasty

This surgical procedure is indicated for bowel-length preservation, especially in Type IIIb atresia and for high jejunal atresias. The bulbous, hypertrophied proximal bowel is derotated and resected back along the antimesenteric border into the third or second part of the duodenum

(**Figure 36.13**). The tapering is performed over a 22–24 Fr catheter to ensure adequate luminal size. An intestinal autostapling instrument may greatly facilitate this procedure. The linear anastomosis is reinforced with interrupted absorbable 5/0 or 6/0 sutures. Tapering can safely be done over an extended 10–20 cm length. Care should be taken not to damage the bile duct and pancreas during back resection and tapering as bowel tends to twist during this procedure.

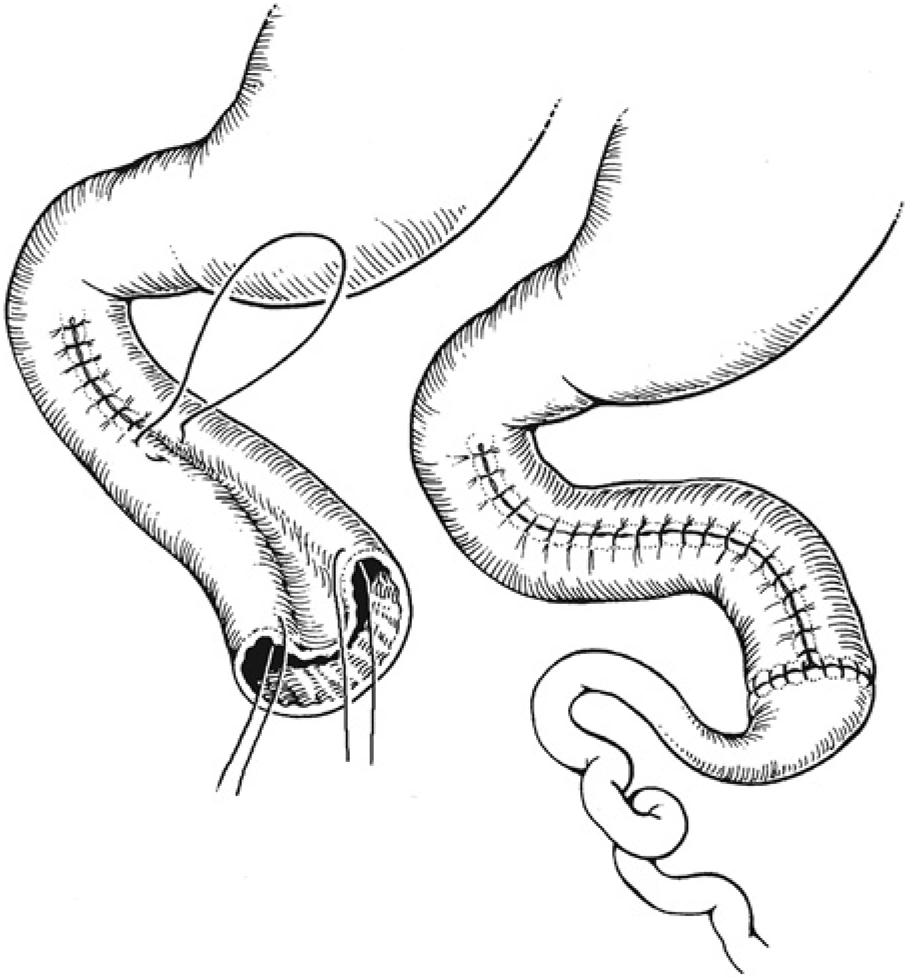


36.13

The tapered bowel is then anastomosed to the distal bowel and replaced in a non-rotated position. Tapering is also indicated for equalizing diameter size for more distal atresias and for correction of a failed inversion plication procedure.

Plication, imbrications, or infolding enteroplasty

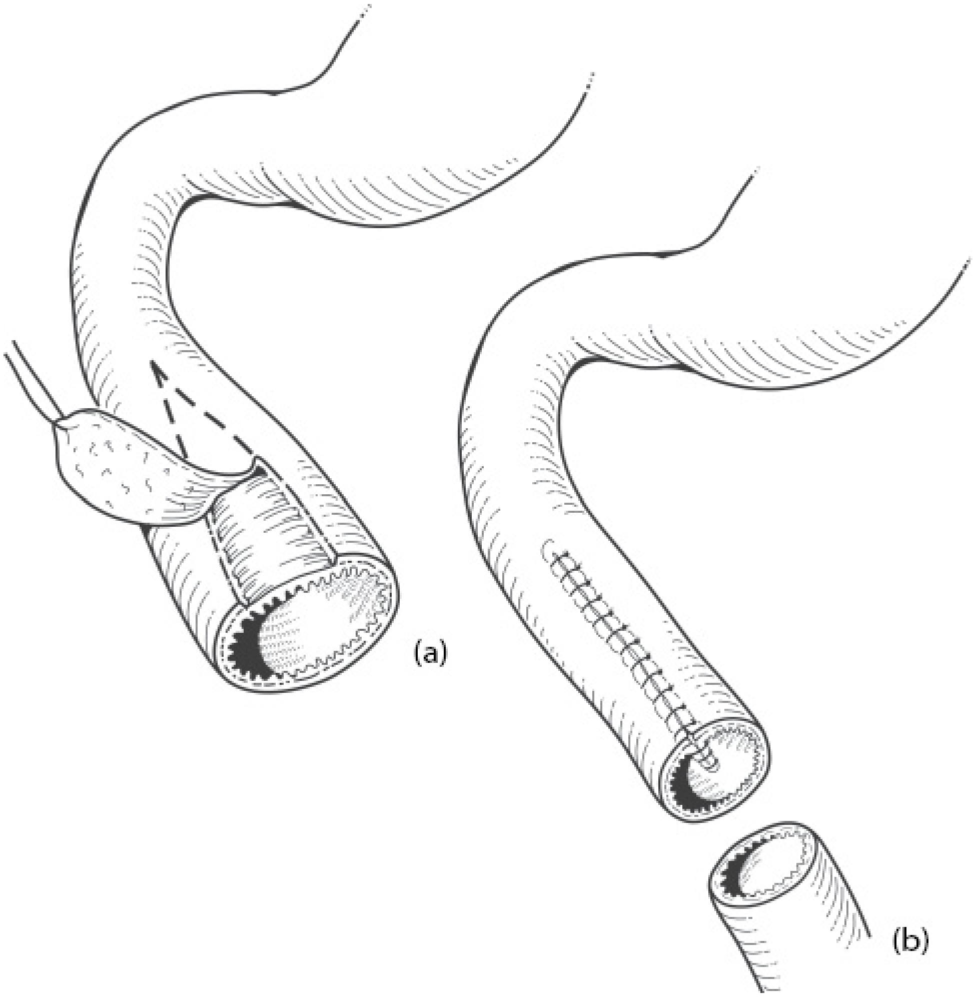
This technique is indicated as for tapering enteroplasty but does not excise any actual mucosa. Antimesenteric intestinal plication involves infolding of up to half or more of the intestinal circumference into the lumen over an extended length using a running stitch (non-absorbable sutures) up to 1 cm from the planned anastomotic site. The distal end is then completed by interrupted stitches to allow for additional surgical trimming if required. The keel created by the in-folding should be sutured closed but avoid compromising luminal patency (**Figure 36.14**).



36.14

Antimesenteric seromuscular stripping and inversion plication

This technique tapers the dilated proximal bowel segment as before but prevents unraveling of a plication enteroplasty. An antimesenteric, seromuscular segment of the dilated proximal bowel is resected by blunt and sharp dissection from the underlying submucosa (**Figure 36.15a**). The muscular margins created are approximated with interrupted sutures, with the underlying mucosa imbricated or inverted into the lumen of the bowel. The distal end of the created keel should be tapered and sutured closed (**Figure 36.15b**). This technique prevents anastomotic leakage and enhances the establishment of prograde peristaltic activity.



36.15a,b

SPECIAL SURGICAL CONSIDERATIONS

Multiple atresias

Multiple atresias may be localized to a short segment of intestine, and resection of several atretic segments with one anastomosis is preferred if sufficient intestinal length remains. If the bowel length is critical, multiple anastomoses should be performed. Placing a transanastomotic intraluminal silastic or PVC tube facilitates the anastomoses.

Bowel lengthening

Bowel-lengthening procedures, such as the serial transverse enteroplasty procedure (STEP), and longitudinal intestinal lengthening and tailoring (LILT) procedures (described by Bianchi) are not recommended at the initial operation (see Chapter 52).

Exteriorization

Exteriorization of proximal and distal intestine may be required with established peritonitis or questionable vascularity of the remaining intestine. The authors do not favor the fashioning of stomas in situations other than these.

Intestinal atresia and gastroschisis

Primary anastomosis may be difficult and hazardous, and the favored option is reduction of the eviscerated intestine with the atresia left intact, closure of the abdominal wall defect, and delayed resection and primary anastomosis of the atresia at 14–21 days.

Colonic atresia

The incidence of colonic atresia is less than small bowel atresia (1 in 66 000) occurring in up to 10% of all atresias. The commonest site is the watershed area between superior mesenteric and inferior mesenteric arterial flow at the distal transverse colon. Prematurity is common and they may be associated with other congenital abnormalities. Colonic atresias may be seen distal to small bowel atresias and also may be associated with conditions such as gastroschisis and Hirschsprung disease. Typically, they present with signs and symptoms of distal bowel obstruction. Plain abdominal radiographs may show dilated intestinal loops with air–fluid levels. If the ileocecal valve is competent, then there is a single, large right-sided "banana-shaped" loop representing the closed loop of dilated ascending and transverse colon. Contrast enema confirms the presence of a microcolon and is very helpful in assessing the level of atresia.

Surgical options include resection and primary anastomosis or initial stoma formation and later closure of the stomas. It is important to establish presence or absence of the distal pathology, such as Hirschsprung disease, if primary anastomosis is contemplated. Most recommend an initial defunctioning colostomy followed by delayed bowel anastomosis, however, primary anastomosis may be feasible in selected cases where the caliber discrepancy is not too marked.

POSTOPERATIVE CARE

Nasogastric decompression is usually required for 4–6 days after the operation (longer for high jejunal atresias). Therapeutic antibiotics are continued for 5–7 days or longer, and an oral antifungal agent is given prophylactically. Oral intake is commenced when the neonate is alert, sucks well, and there is evidence of prograde gastrointestinal function, i.e. clear gastric effluent of low volume, a soft abdomen, or when flatus or feces have been passed. Surveillance should continue until the infant has established normal gastrointestinal function.

If at any time there is suspicion of a leak at the anastomosis (suggested by ileus, abdominal distension, vomiting, and peritonitis), a plain erect or decubitus radiograph of the abdomen should be taken. If this reveals free air in the abdomen more than 24 hours after operation, laparotomy should be performed and the leaking site resutured or revised.

COMPLICATIONS

Although a survival rate of >90% can be expected in well-resourced centers, complications are not infrequent. These include anastomotic leaks and stricture formation, ischemia of the bowel due to a precarious blood supply especially in Type IIIb atresias, adhesive obstruction, short bowel syndrome, and infections related to pneumonia and septicemia.

OUTCOME IN RED CROSS HOSPITAL, CAPE TOWN, SOUTH AFRICA

Before 1952, the mortality rate for congenital atresias of the small intestine was 90%. This diminished to 70% between 1952 and 1955. At that stage, most were treated by primary anastomosis without resection. With liberal back resection of the proximal blind end and end-to-end anastomosis, the survival rate increased to 80% from 1955 to 1958.

During the 59-year period 1959–2017, 351 patients with jejunoileal atresia and stenosis were admitted to the Pediatric Surgical Service at the Red Cross Children's Hospital, Cape Town, South Africa, of whom 38 died, giving an overall mortality rate of 10.8% (**Tables 36.1–36.3**). However, in line with the improved preoperative and postoperative care of newborns, our last 21-year mortality rate dropped to 6% (nine deaths in 148 patients). The causes of death in these nine infants were extreme short bowel syndrome (n = 5), moribund at presentation due to volvulus and necrotic bowel (Type IIIb) (n = 1), congenital syphilis (n = 1), HIV infection (n = 1), sepsis (n = 1), and cot death at 2 months of age (n = 1).

Table 36.1 Jejunal atresia and stenosis (n = 267) and ileal atresia and stenosis (n = 84): Experience at Red Cross Children's Hospital, Cape Town, South Africa, 1959–2017

Type	Jejunum	Ileum	Total	(%)
Stenosis	21	13	34	9.69
Type 1	64	20	84	23.93
Type 2	20	13	33	9.40
Type 3a	28	24	52	14.81
Type 3b	65	0	65	18.52
Type 4	69	14	83	23.65
Total	**267**	**84**	**351**	**100.00**

Table 36.2 Mortality related to type of atresia at Red Cross Children's Hospital, Cape Town, South Africa, 1959–2017

Type	Patients	Mortality	%
Stenosis	34	0	0
Type 1	84	4	4.8
Type 2	33	4	12.1
Type 3a	52	8	15.4
Type 3b	65	12	18.5
Type 4	83	10	12.1
Total	**351**	**38**	**10.8**

Table 36.3 International experience of jejunoileal atresia and stenosis: Improvement in survival

Authors	City/Country	Years of study	*n*	Survival (%)
Evans		1950	1498	9.3
Gross	Boston, USA	1940–1952	71	51
Benson et al.	USA	1945–1959	38	55
De Lorimier	Los Angeles, USA	1957–1966	587	65
Nixon and Tawes	London, UK	1956–1967	62	62
Louw	Cape Town, South Africa	1959–1967	33	94
Martin and Zerella	USA	1957–1975	59	64
Cywes et al.	Cape Town, South Africa	1959–1978	84	88
Smith and Glasson	Camperdown, Australia	1961–1986	84	61
Vecchia et al.		1972–1997	128	84
Millar et al.	Cape Town, South Africa	1990–2011	133	95

FURTHER READING

Benson CD, Lloyd JR, Smith JD. Resection and primary anastomosis in the management of stenosis and atresia of the jejunum and ileum. *Pediatrics* 1960; 26: 265–72.

Kimura K, Perdzynski W, Soper RT. Elliptical resection of tapering the proximal dilated bowel in duodenal or jejunal atresia. *J Pediatr Surg* 1996; 31: 1405–6.

Kling K, Applebaum H, Dunn J, et al. A novel technique for correction of intestinal atresia at the ligament of Treitz. *J Pediatr Surg* 2000; 35: 353–6.

Louw JH, Barnard CN. Congenital intestinal atresia. *Lancet* 1955; 269: 1065–7.

Smith GHH, Glasson M. Intestinal atresia: Factors affecting survival. *ANZ J Surg* 1989; 59: 151–6.

Spencer AU, Neaga A, West B, et al. Pediatric short bowel syndrome redefining predictors of success. *Ann Surg* 2005; 242: 403–12.

Touloukian RJ, Walker Smith GJ. Normal intestinal length in preterm infants. *J Pediatr Surg* 1983; 18: 720–3.

37

Meconium ileus

ROBERT J. VANDEWALLE AND FREDERICK J. RESCORLA

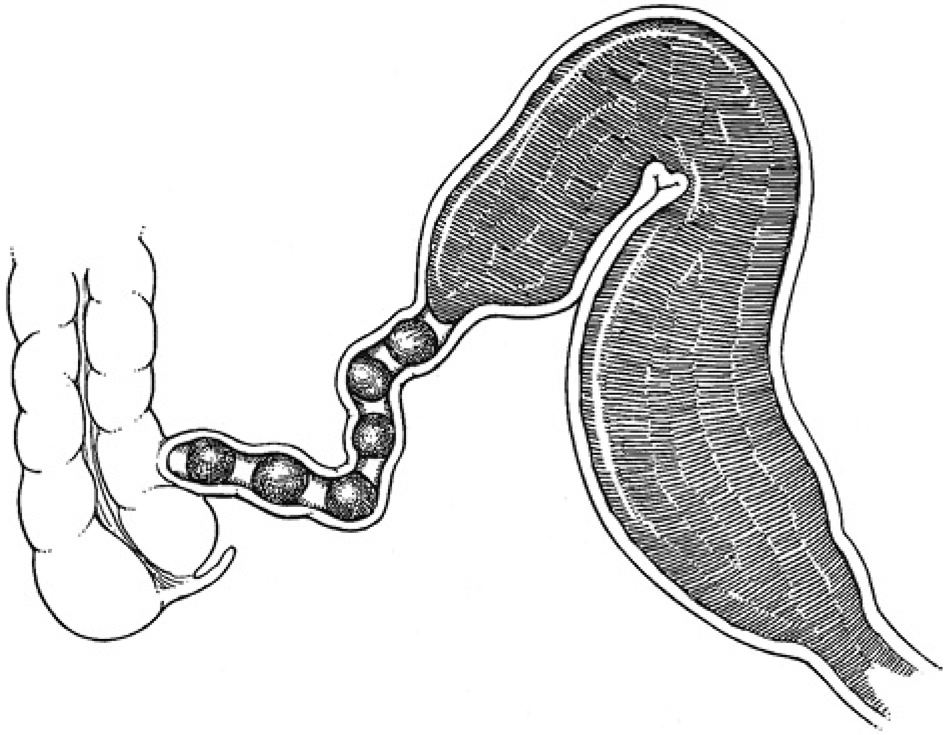
ETIOLOGY AND CLASSIFICATION

Cystic fibrosis (CF) is the most common serious inherited defect affecting the Caucasian population. CF is transmitted as an autosomal recessive condition with a 5% carrier rate and an incidence of approximately 1 in 2500 live births. The CF transmembrane conductance regulator (*CFTR*) gene is located on the long arm of chromosome 7. According to the Cystic Fibrosis Genetic Analysis Consortium, 13 mutations occur at a frequency of greater than 1% and account for 87% of CF alleles. The delta F508 mutation is the most common and is present in 70% of CF alleles in the United States. There are great differences among populations, and among African Americans delta F508 accounts for only 43% of the alleles. The CFTR protein controls sodium and chloride transport and in CF this results in abnormal luminal secretions. Neonatal intestinal obstruction due to inspissated meconium has been identified since the early reports concerning CF and is referred to as meconium ileus. This presentation is observed in up to 20% of infants born with CF. The etiology of this abnormal meconium (mucoviscidosis) is due to deficient pancreatic and intestinal secretions, as well as an abnormal concentration of the meconium within the duodenum and proximal jejunum. Instances of meconium ileus can be classified into uncomplicated and complicated cases.

Uncomplicated meconium ileus

CLINICAL PRESENTATION

In uncomplicated meconium ileus, the abnormal thickened meconium causes a simple obturator obstruction of the terminal ileum. The distal 15–30 cm of terminal ileum is filled with inspissated meconium pellets, which are adherent to the bowel wall. The ileum just proximal to the obstruction fills with thick, putty-like meconium and dilates to 3–4 cm in diameter (**Figure 37.1**). The colon is unused and small (microcolon), because meconium has not yet entered this segment of bowel.

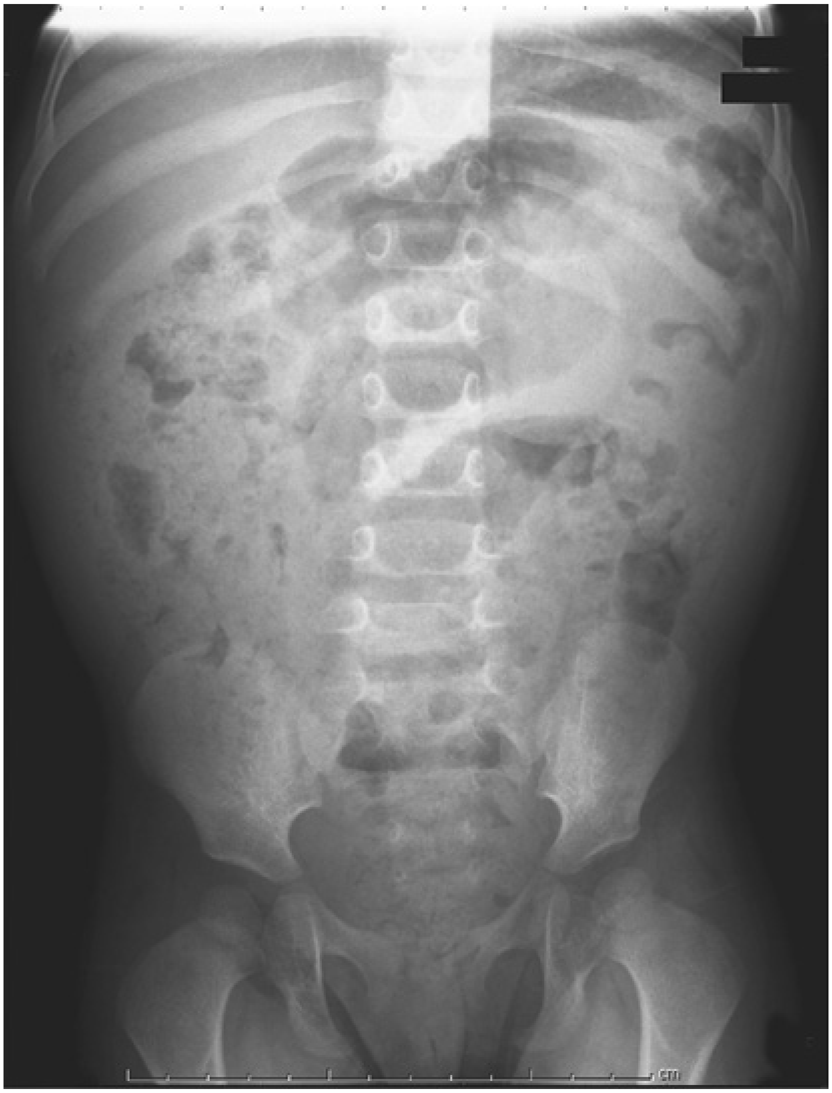


37.1

The typical neonate with meconium ileus may appear relatively normal for the first 12–18 hours of life and some tolerate several feeds. As the proximal bowel fills with swallowed air, however, abdominal distension and emesis (initially clear, later bilious) and failure to pass meconium are noted, heralding the presence of intestinal obstruction at 24–36 hours of age.

RADIOGRAPHIC EVALUATION

Plain abdominal X-rays and decubitus views usually demonstrate similar-sized dilated loops of intestine without air–fluid levels. A “soap bubble” appearance is often noted in the right lower quadrant, a result of air mixing with the thick meconium (**Figure 37.2**).



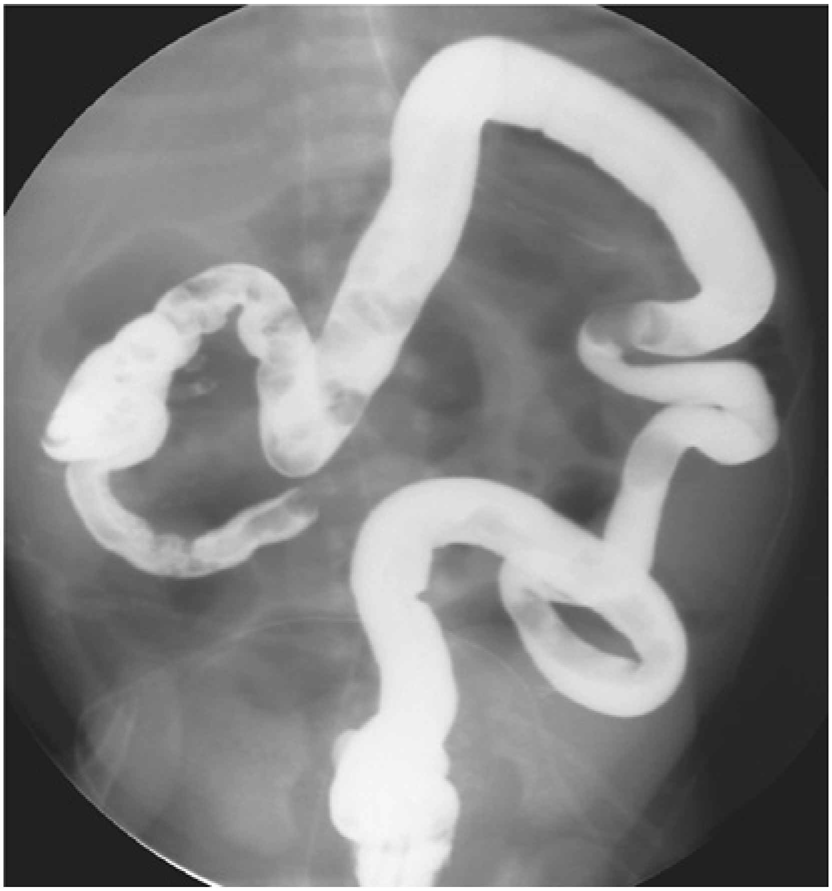
37.2

INITIAL MANAGEMENT

The neonate with suspected bowel obstruction should be treated with oral gastric tube decompression of the stomach and IV fluids to replace pre-existing fluid deficits and ongoing losses. Antibiotics are administered, as the differential diagnosis of a newborn with this presentation includes sepsis.

DIAGNOSTIC ENEMA

The initial diagnostic test is a contrast enema. If the diagnosis of meconium ileus is not apparent from the plain X-rays, a barium enema is the diagnostic procedure of choice. This will demonstrate a microcolon and may also document the presence of meconium pellets in the proximal ascending colon and terminal ileum (**Figure 37.3**).



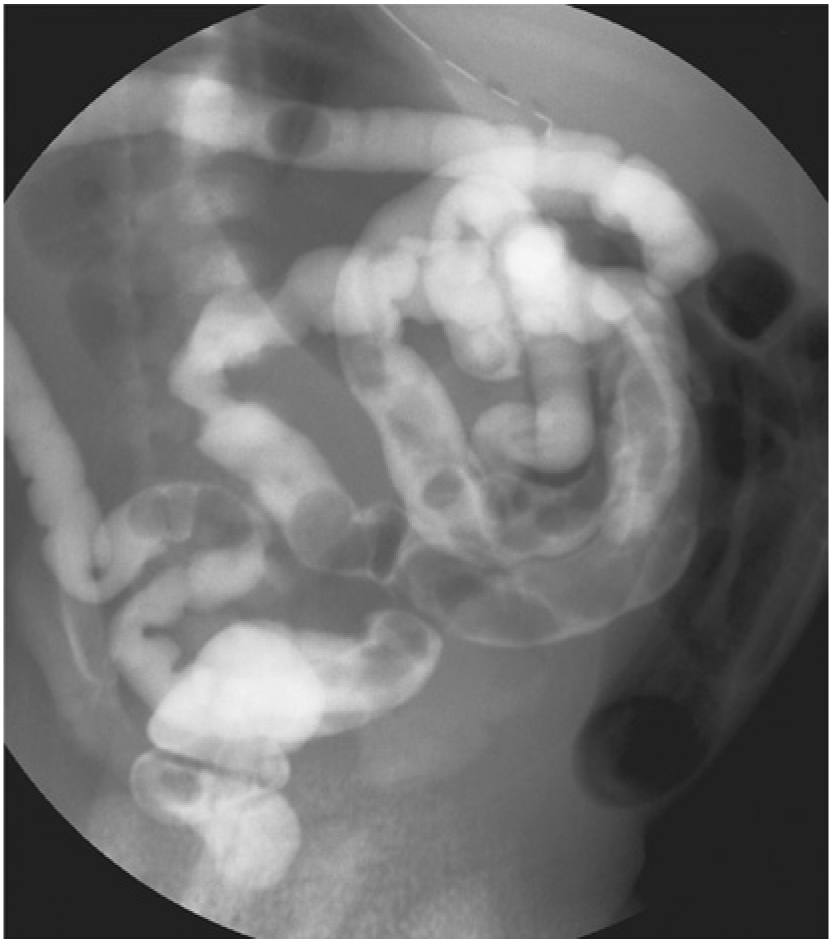
37.3

This study will also exclude cases of colon atresia, small left colon syndrome, and meconium plug syndrome, and document the location of the cecum to rule out anomalies of rotation and fixation. Neonates with distal ileal atresia and total colonic aganglionosis may have similar appearance on a contrast enema examination, but these neonates usually have air–fluid levels in the dilated proximal small bowel and absence of pellets in the distal ileum and proximal colon. If the neonate is stable and there is no evidence of complicated meconium ileus (e.g. peritoneal calcifications, giant cystic structure), an attempt at non-operative treatment with a hypertonic contrast material enema is recommended.

THERAPEUTIC ENEMA

The management of neonates with uncomplicated meconium ileus was significantly altered with the introduction of the diatrizoate meglumine (Gastrografin) enema by Noblett in 1969. The efficacy of this procedure is related to the hyperosmolar nature of Gastrografin (1100–1900 mOsm/L), which draws large volumes of fluid into the bowel lumen, thus washing out the obstructing meconium. Currently, most pediatric radiologists dilute the contrast material to approximately 3 : 1. Complications reported following Gastrografin enema include perforation, necrotizing enterocolitis, shock, and the occasional death. Most of these events are probably related to the hyperosmolar nature of the contrast material causing fluid depletion, which results in decreased intestinal blood flow and perfusion. Some radiologists prefer using agents such as diatrizoate sodium (Hypaque) or iothalamate meglumine (Conray), alone or in combination with *N*-acetylcysteine, while the authors prefer to use dilute Gastrografin. It is essential that the radiologists and clinicians are aware of the osmolality of the solution.

The enema is gently administered under fluoroscopic control and the contrast material flushed around the obstructing meconium pellets in the terminal ileum (**Figure 37.4**).



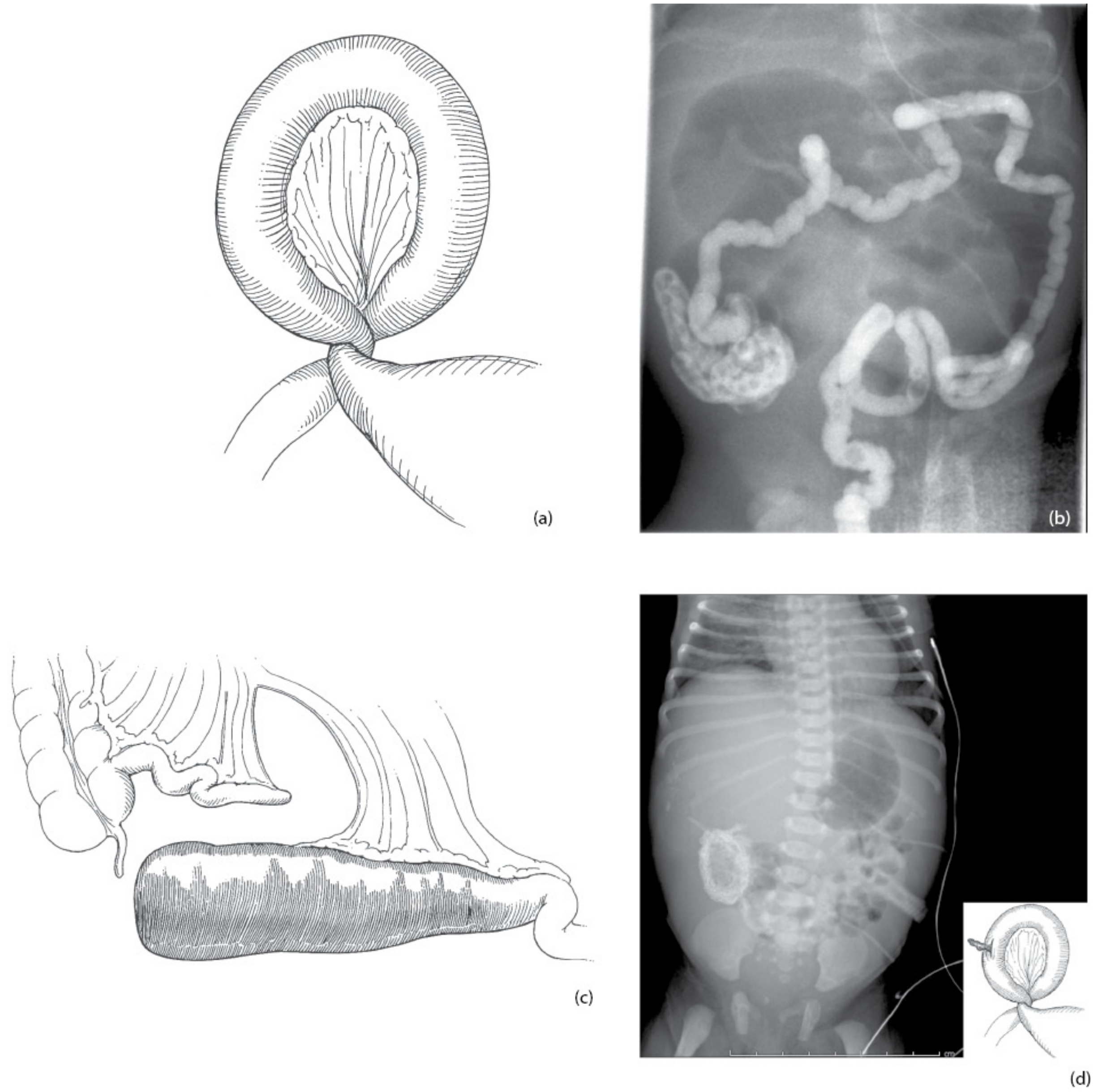
37.4

Before the enema the infant must be appropriately resuscitated. Intravenous fluids are infused at a rate of 1.5 times maintenance during and after the procedure. The infant's pulse rate and urine output are carefully monitored in anticipation of fluid shifts. Meconium pellets, followed by loose meconium, generally pass through the rectum over the next 4–8 hours. If evidence of bowel obstruction persists and the infant remains stable, a second or third enema may be administered. *N*-acetylcysteine (2.5–5%, 5 mL every 6 hours) may be administered by oral gastric tube to aid clearing the thickened meconium. As the clinical evidence of obstruction resolves, the oral gastric tube is removed and feeding advanced. The Gastrografin enema is successful in resolving the obstruction in approximately 55% of cases. Survival for these infants at one year is nearly 100%. If these non-operative efforts fail, surgical exploration is required.

Complicated meconium ileus

CLINICAL PRESENTATION AND INITIAL MANAGEMENT

Complicated cases include instances of volvulus, bowel perforation, intestinal atresia, and giant cystic meconium peritonitis. Volvulus usually occurs when the distended segment of ileum twists at the level of the narrow, pellet-filled, distal small intestine (**Figure 37.5a**). In some cases, volvulus can result in bowel perforation, leading to meconium peritonitis, and, in others, the bowel may become necrotic and liquefy, resulting in a pseudocyst (i.e. giant cystic meconium peritonitis (**Figure 37.5b**)). Bowel atresias are thought to arise when the base of the volvulus becomes ischemic (**Figure 37.5c**). Contrast enema studies will not reflux into the dilated segment of small intestine in any of these scenarios (**Figure 37.5d**).

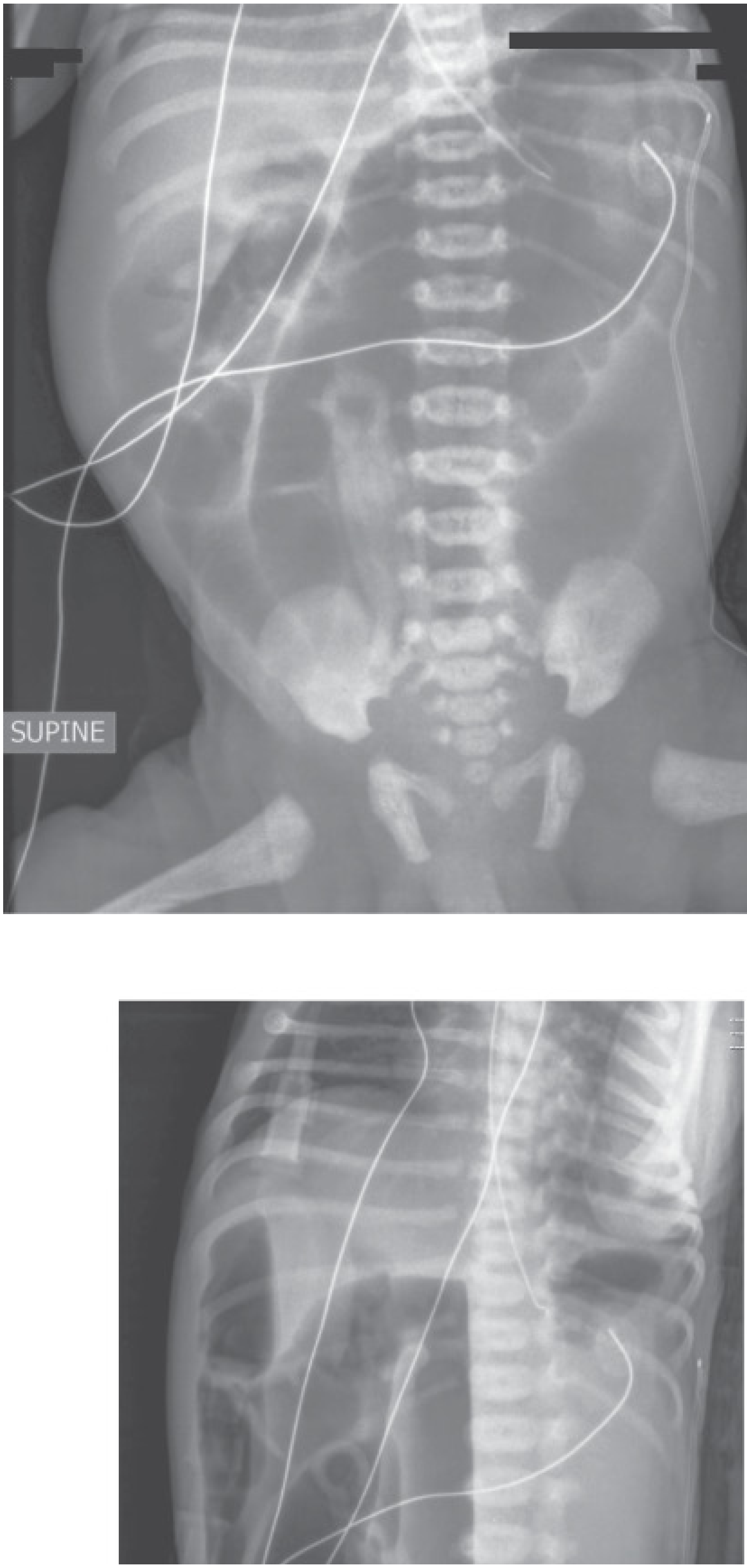


37.5a–d

Neonates with complicated meconium ileus usually present with abdominal distension at the time of, or shortly after, delivery. In addition, bile-stained fluid is usually noted in the stomach. On physical examination, an abdominal mass may be noted. Neonates with meconium peritonitis occasionally have meconium in the scrotal sac or vagina as a result of passage of this material through a patent processus vaginalis or the fimbriated ends of the Fallopian tubes, respectively. In one unusual report, a meconium pseudocyst appeared as a buttock mass. The early management of these neonates includes IV hydration, antibiotics, and oral gastric tube decompression of the stomach.

DIAGNOSIS

In contrast to neonates with uncomplicated meconium ileus, flat and erect or decubitus X-rays of the abdomen in complicated cases may demonstrate distended loops of small bowel of different size with air–fluid levels (**Figure 37.6a**). Intraperitoneal calcifications from the extravasated meconium, characteristic of meconium peritonitis, may be noted (**Figure 37.6b**). A mass effect or ascites may also be observed.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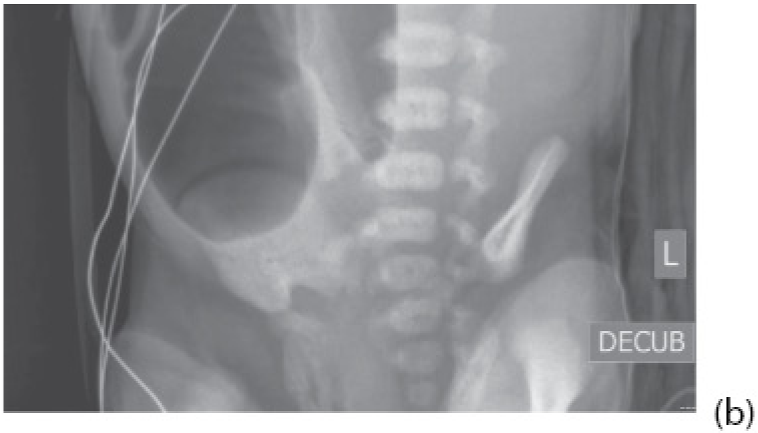


37.6a,b

Neonates who can be identified as complicated cases by plain abdominal radiographs are taken to the operating room for prompt exploration. In uncertain cases, a barium enema may be useful to exclude other causes of distal obstruction.

OPERATIONS

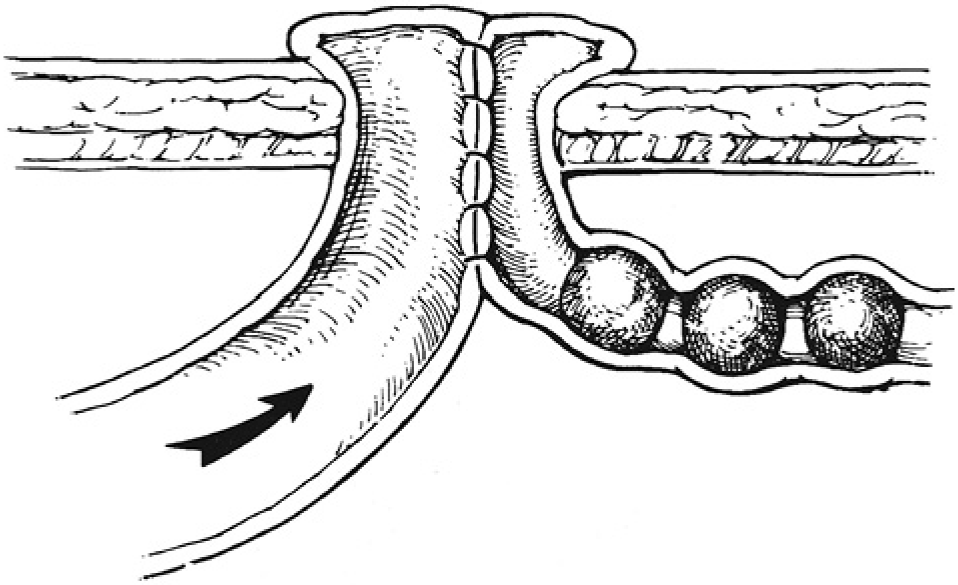
Uncomplicated meconium ileus

MIKULICZ PROCEDURE

Meconium ileus was often considered a fatal condition until 1948 when Hiatt and Wilson reported a number of survivors after enterotomy and irrigation. This technique was not widely utilized and, in 1953, Gross reported successful outcomes in infants with meconium ileus following bowel resection and use of Mikulicz enterostomy.

The dilated bowel loop filled with thickened meconium is brought out of the abdomen and the small bowel proximal and distal to this segment is sutured together in a side-to side fashion by interrupted seromuscular sutures (**Figure 37.7**). Following closure of the abdomen, the exteriorized dilated bowel is resected, thus avoiding the risk of peritoneal contamination. The distal bowel can be irrigated in the postoperative period to wash out the obstructing meconium pellets through the enterostomy. A

Mikulicz spur-crushing clamp is applied, resulting in a common lumen, and the ostomy is then closed at a later date.

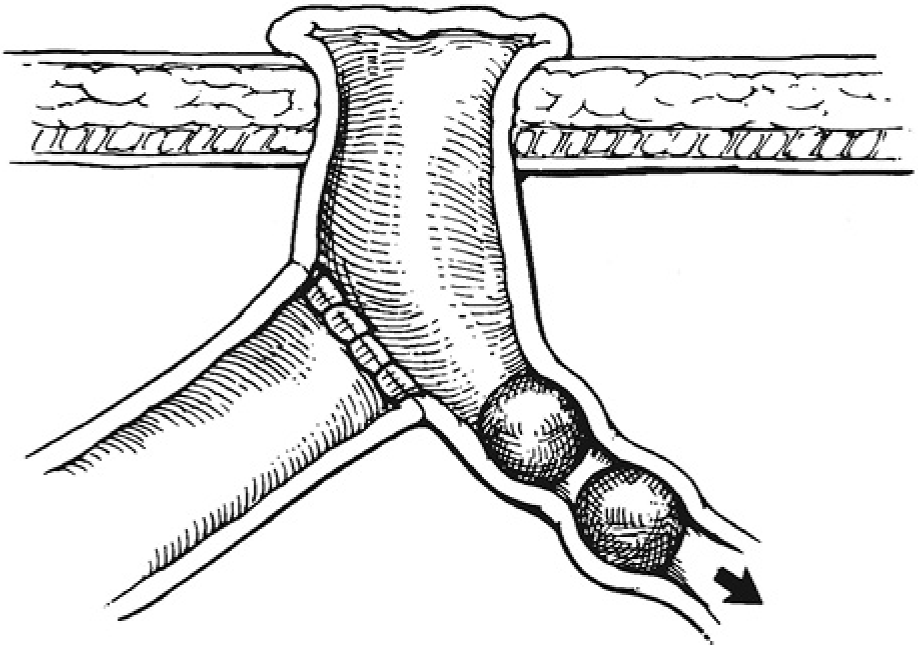


37.7

The disadvantages of this procedure are the loss of fluids from the mid-small bowel ostomy, the need for a subsequent procedure to close the stoma, and some reduction of bowel length due to initial resection.

BISHOP–KOOP PROCEDURE

In 1957, Bishop and Koop reported resection of the large dilated loop followed by an anastomosis between the end of the proximal segment and the side of the distal segment with the distal bowel brought out as an end ileostomy (**Figure 37.8**). A catheter is passed into the distal segment to allow postoperative irrigation. As the distal obstruction is relieved, the intestinal contents preferentially pass into the distal ileum and colon, thus decreasing loss of fluid and electrolytes from the stoma. The ostomy can be closed at a later date and, in some cases where it is trimmed beneath the skin, may close spontaneously.

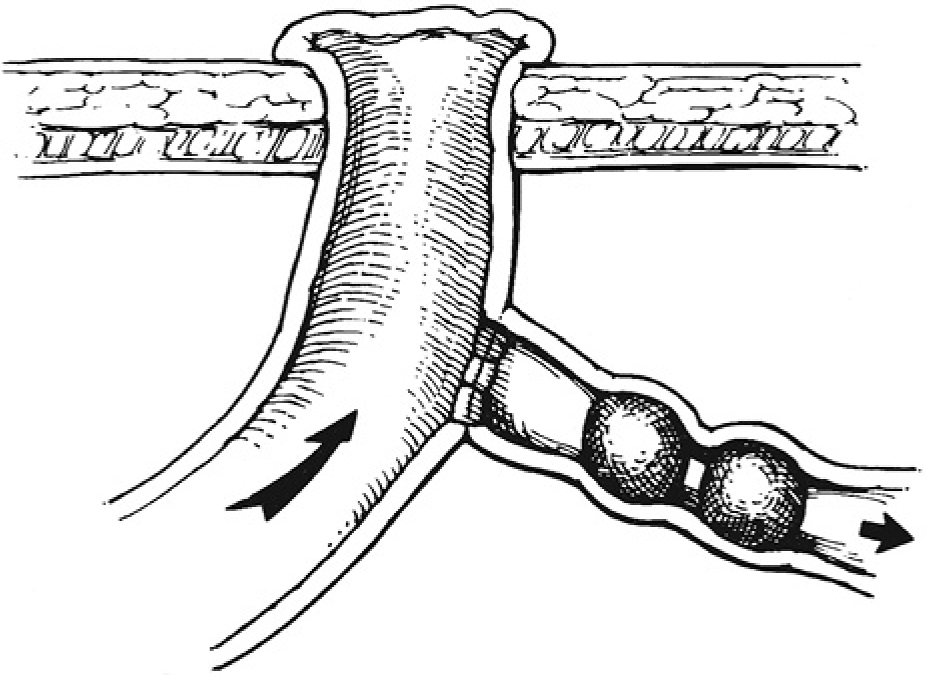


37.8

The disadvantages of this technique include loss of bowel length at the time of the initial procedure, the need for an intraperitoneal anastomosis, and the need for a second operative procedure.

SANTULLI–BLANC ENTEROSTOMY

This modification of the Bishop–Koop procedure concept was reported in 1961. The operation involves resection of the distal dilated bowel segment followed by a side-to-end anastomosis with proximal enterostomy (**Figure 37.9**).



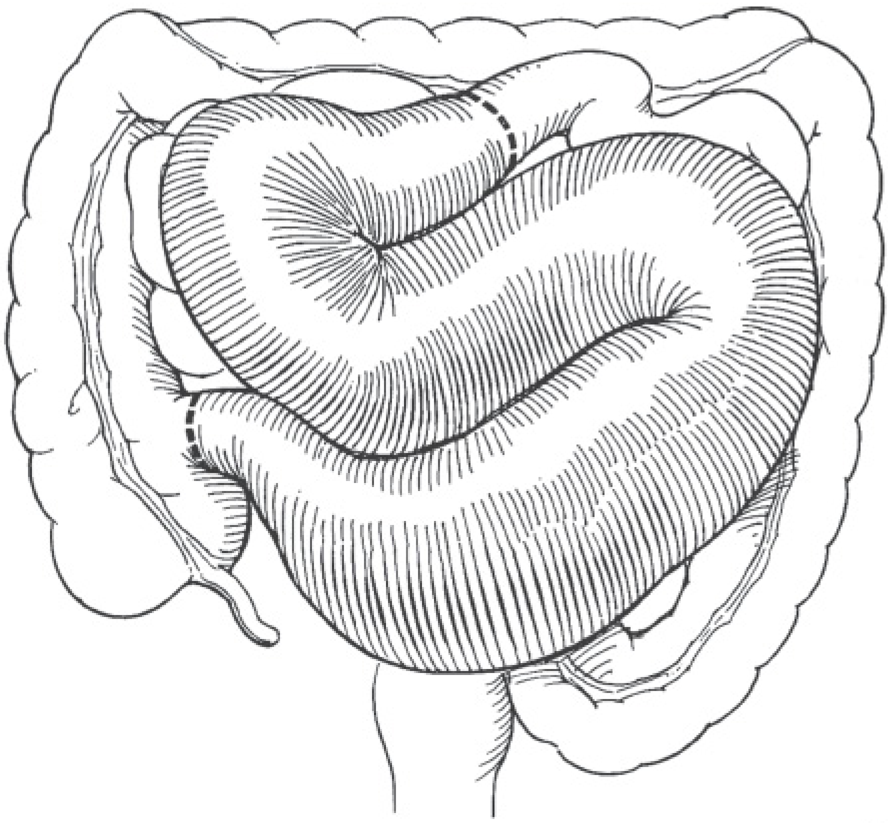
37.9

The disadvantages are similar to those noted for the Bishop–Koop procedure.

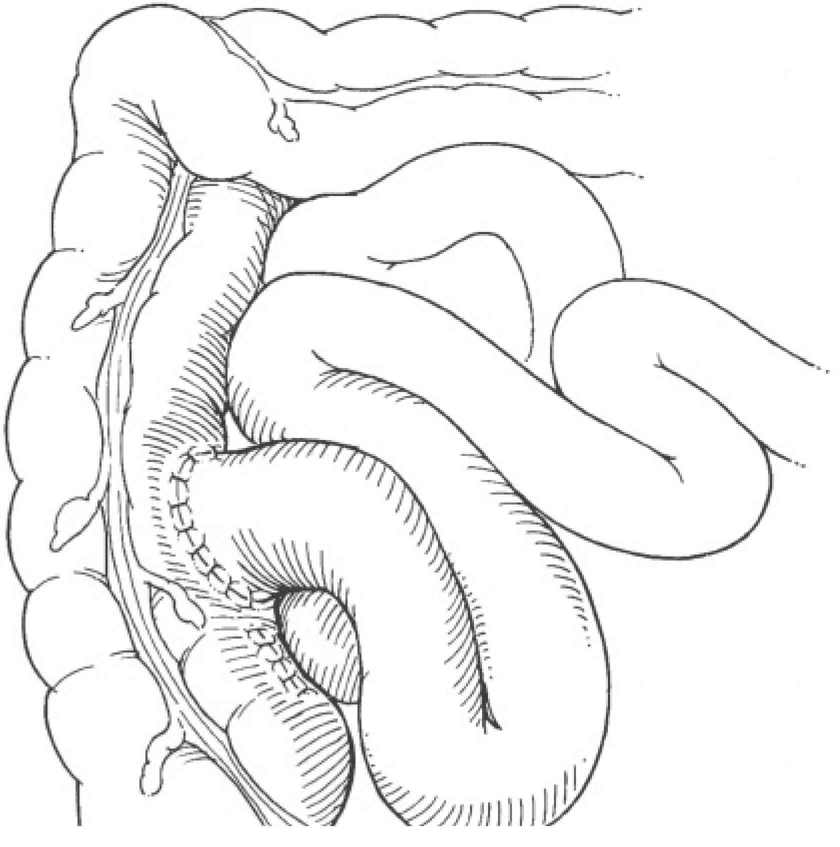
PRIMARY RESECTION AND ANASTOMOSIS

The use of resection with primary anastomosis in the management of meconium ileus was first reported by Swenson in 1962.

After resection of the obstructed bowel segment, the remaining pellets in the distal bowel are irrigated clear and an ileocolonic anastomosis is performed (**Figure 37.10a and b**).



(a)

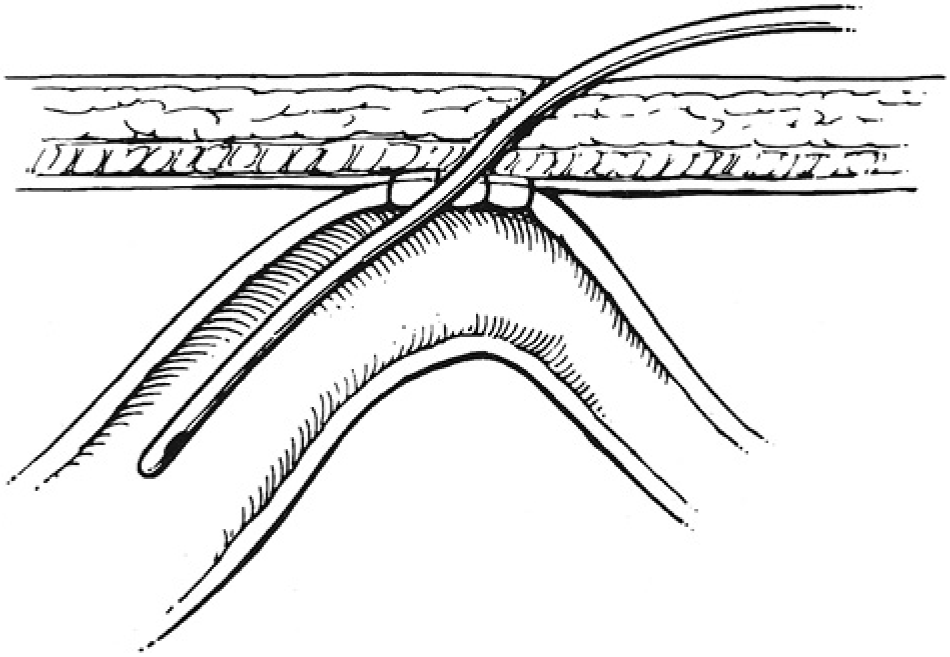


37.10a,b

The disadvantage associated with this procedure is resection of additional bowel, as the terminal ileum containing meconium pellets was usually resected along with the dilated segment of ileum. This, as well as concerns about an unvented intraperitoneal anastomosis, has prevented wide acceptance of this procedure.

TUBE ENTEROSTOMY

In 1970, O'Neill et al. reported success with a simple procedure involving tube enterostomy with postoperative irrigation (**Figure 37.11**). Their initial report of five neonates was followed by a report by Harberg et al. concerning 9 of 11 neonates who had successful meconium washout using this technique. Harberg et al. modified the technique slightly by utilizing a T-tube. A follow-up study of this technique noted success in 20 out of 23 patients with T-tube irrigation utilizing pancreatic enzymes or 1% *N*-acetylcysteine, 5–12.5 mL per dose. Unfortunately, three patients required a second procedure in the neonatal period due to inadequate treatment with the T-tube.

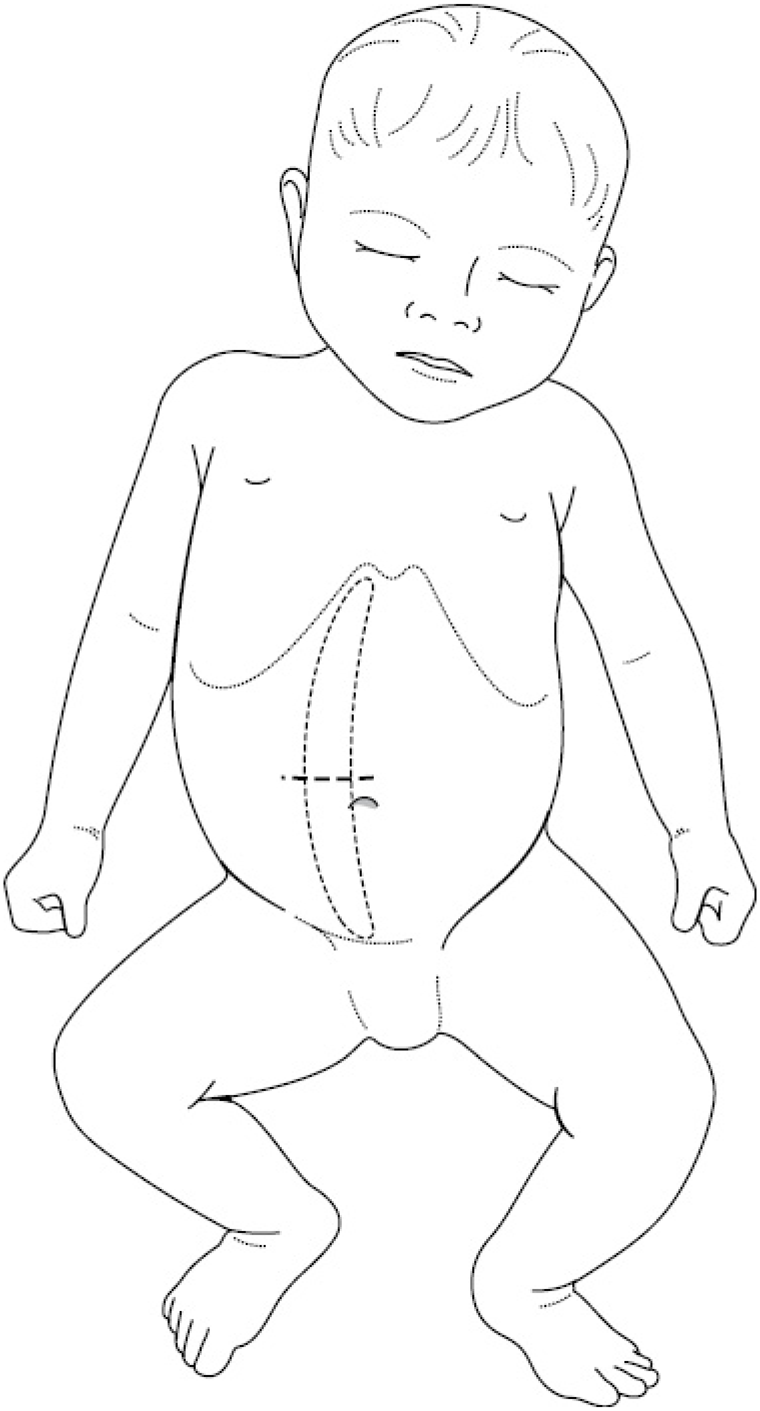


37.11

ENTEROTOMY AND IRRIGATION

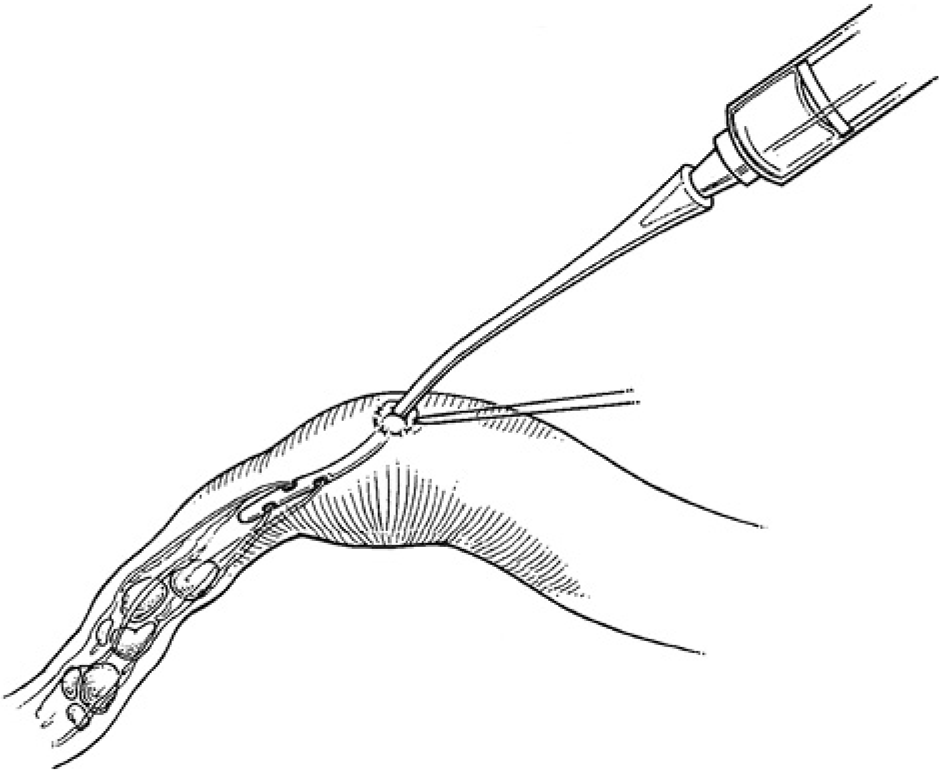
The technique of enterotomy with irrigation has been one the procedures of choice at our institution since 1981. As previously noted, this procedure was originally described by Hiatt and Wilson in 1948. Reports by several other authors appeared in the literature between 1970 and 1990.

After induction of general anesthesia, the procedure is carried out through a right-sided supraumbilical transverse abdominal incision (**Figure 37.12**). The right rectus abdominis muscle, as well as a portion of the right external and internal oblique and transversus abdominis muscles, is divided using electrocautery. The peritoneal cavity is entered and explored. The distended meconium-filled loops and pellet-filled distal small bowel are identified and delivered into the wound.



37.12

A purse-string suture of 4/0 silk is placed on the antimesenteric border of the dilated bowel 6–8 cm proximal to the narrow region containing the obstructing pellets. An 8–10 Fr red rubber catheter is placed through a small enterotomy, the purse-string suture is snugged, and the bowel lumen is irrigated by gently instilling saline through a syringe attached to an adapter and three-way stopcock. Fluid is irrigated into the proximal thick meconium and around the distal pellets (**Figure 37.13**).

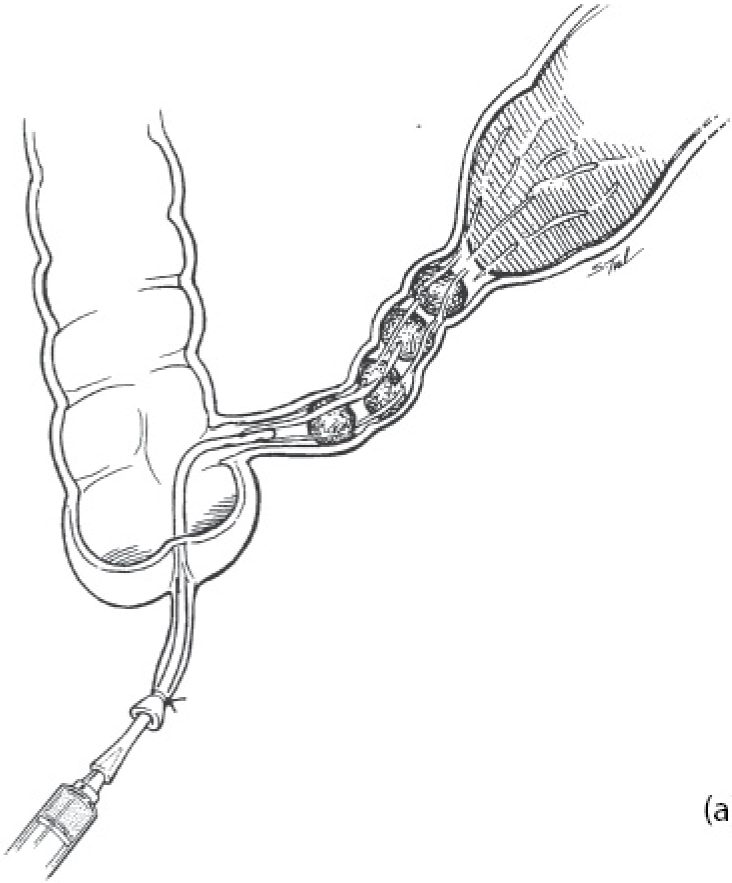


37.13

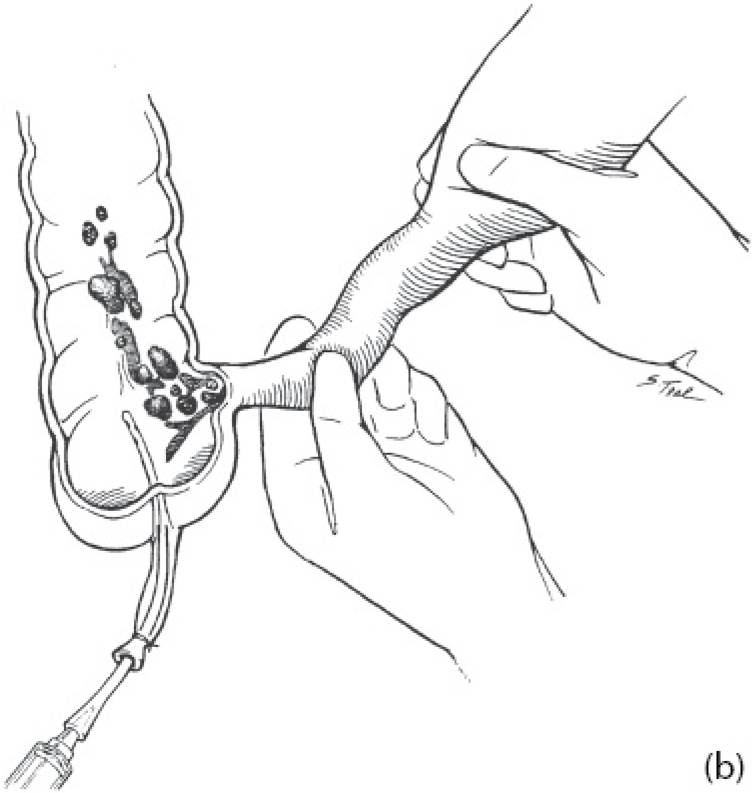
With irrigation and gentle manual manipulation, the thick meconium from the distended loop and the distal pellets can be removed through the enterotomy or washed out distally through the colon. Occasionally, the enterotomy may need to be extended to a 1.5–2.0 cm opening to allow removal of the thick meconium and pellets. Numerous irrigations are usually required. Dilute Gastrografin or 2.5–5% *N*-acetylcysteine solution may also be used for irrigation. There have been occasional reports documenting hypernatremia with *N*-acetylcysteine. When the pellets are cleared, saline is irrigated through the distal ileum and into the colon to exclude the possibility of atresia and also to flush some of the distal pellet fragments into the colon. The enterotomy is then closed in two layers, using an inner full-thickness layer of absorbable 4/0 or 5/0 polyglactin (running or interrupted) and an outer seromuscular layer with interrupted 5/0 silk suture.

APPENDICAL IRRIGATION

In 1989, Fitzgerald and Conlon reported the use of the appendix to instill dilute Gastrografin around the thickened meconium and pellets and flush the material into the colon. The catheter can usually be advanced retrogradely into the terminal ileum, and saline is gently irrigated (**Figure 37.14a**); the surgeon gently compresses the pellets and meconium to allow smaller particles to mix with the saline. The catheter is then withdrawn into the appendix and the surgeon "milks" the material into the colon (**Figure 37.14b**). This process must usually be repeated several times and, as the material is advanced into the ascending colon, it is gently flushed into the distal colon and out of the rectum. After the ileal obstruction is completely relieved, an appendectomy is performed. We have found this difficult to accomplish as the thickened meconium in the terminal ileum impedes passage of the catheter and mobilization of the pellets.



(a)



(b)

37.14a,b

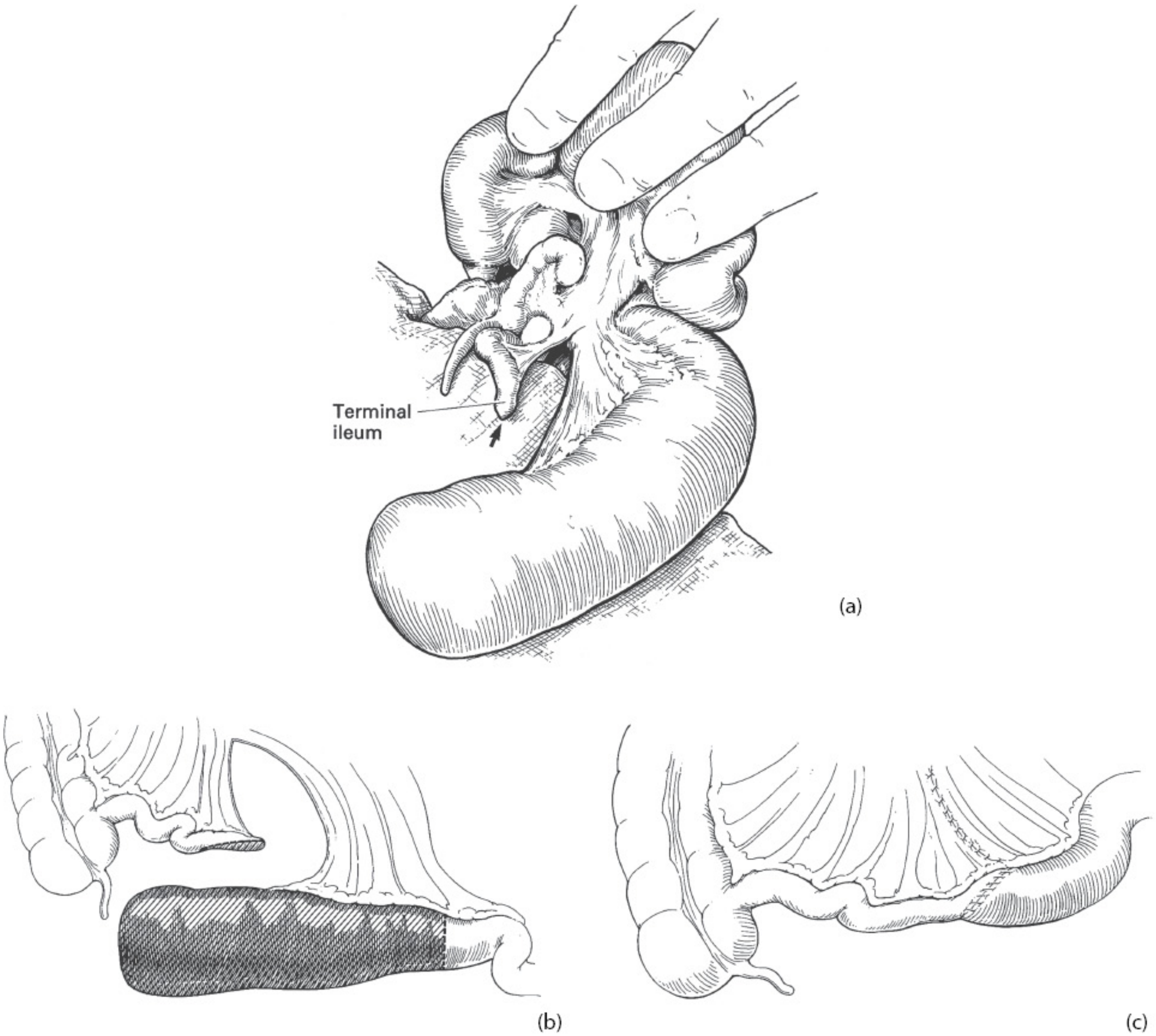
Complicated meconium ileus

The anesthetic management is similar to that described for uncomplicated cases. The abdomen is entered through a transverse supraumbilical incision. The operative findings and ease of procedure may vary significantly, from volvulus and bowel atresia to meconium peritonitis or giant cystic meconium peritonitis. These conditions will, therefore, be discussed separately.

VOLVULUS AND ATRESIA

In instances of meconium ileus associated with bowel volvulus or atresia, the pathology is usually easily identified at laparotomy, and a primary anastomosis is nearly always possible.

In cases of volvulus, the involved loop is resected and an end-to-oblique anastomosis is constructed between the dilated proximal bowel and smaller distal bowel. The proximal bowel is divided at a 90-degree angle with respect to the mesentery and the distal bowel at a 45-degree angle (**Figure 37.15a**). The distal meconium pellets should be removed through the open bowel, and the distal segment should also be irrigated to facilitate return of bowel function and to avoid a postoperative obturator obstruction distal to the anastomosis (**Figure 37.15b**). The anastomosis is constructed with one or two layers of interrupted 5/0 silk sutures (**Figure 37.15c**).



37.15a–c

In cases of atresia, if adequate bowel length is present, the proximal dilated segment (usually 10–15 cm) is resected, as it is frequently atonic, and an end-to-oblique anastomosis is fashioned.

MECONIUM PERITONITIS AND GIANT CYSTIC MECONIUM PERITONITIS

Neonates with these two disorders generally have numerous adhesions throughout the abdomen and may have significant blood and fluid losses during the operation. The abdomen is explored and the small bowel and colon carefully identified and dissected free from the numerous adhesions.

The necrotic dilated segment is resected and, if possible, an end-to-end or end-to-oblique anastomosis is constructed. In most cases, this is not possible and a temporary enterostomy is required. The bowel opening may be found within the pseudocyst. Most of the pseudocyst wall should be resected, if possible. This may result in some blood loss requiring transfusion. The proximal end may be brought out as a temporary stoma through the corner of the wound or through a separate incision. The distal bowel can either be closed with sutures or staples and left in the abdomen, or brought out as a separate mucous fistula to allow irrigation, as well as refeeding of the effluent from the proximal ostomy into the distal bowel. In 1993, Tanaka et al. reported treating two cases of cystic meconium peritonitis with initial ultrasound-guided drain placement followed by delayed, single-stage surgery. The authors reported successful anastomosis and cyst resection with decreased blood loss. While this technique appears promising, our institution has no experience with this method.

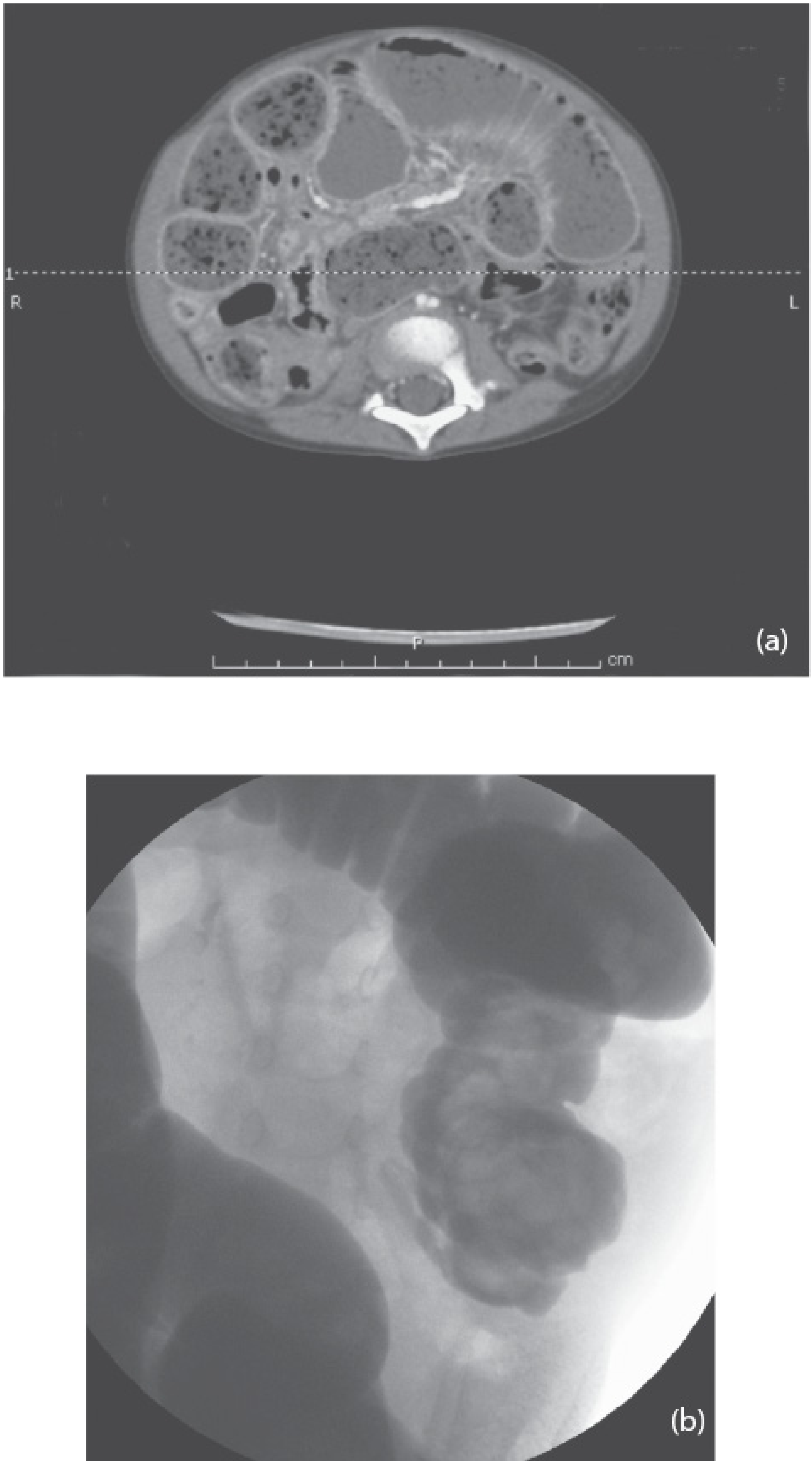
POSTOPERATIVE CARE

Uncomplicated meconium ileus

In the early postoperative period, an oral gastric tube is left in place until bowel function returns. Use of *N*-acetylcysteine (2.5–5%, 5–10 mL in 6 hours) through an oral gastric tube may further aid passage of inspissated meconium. When bowel function returns, the tube is removed and enteral feedings are initiated along with pancreatic enzyme supplementation. The diagnosis of CF is confirmed by obtaining an elevated sweat chloride level on testing and the actual chromosomal defect is identified to assist in genetic counseling. The management of these patients requires a multidisciplinary team, including the pediatric pulmonary physician, in order to optimize the pulmonary status, as pulmonary function deterioration is the major cause of morbidity and mortality.

Distal intestinal obstruction syndrome

The clinical presentation of distal intestinal obstruction syndrome, formerly referred to as meconium ileus equivalent, includes abdominal pain and decreased stool frequency. Plain X-rays frequently demonstrate large amounts of fecal material with the “bubbly” granular appearance similar to meconium ileus (**Figure 37.16a**). This may require treatment with Gastrografin enemas or administration of a balanced intestinal lavage solution orally or through a nasogastric tube. The obstruction usually responds to non-operative management with a balanced salt solution such as GoLytley (20 mL/kg per hour for 3–4 hours). If dehydration is present, IV hydration will be required. In some more advanced cases or if the diagnosis is not clear, a water-soluble contrast enema with Gastrografin or other solution may be both diagnostic and therapeutic (**Figure 37.16b**). The use of oral or nasogastric Gastrografin has also been reported with a dose of 50 mL followed by at least 200 mL of water in children less than 8 years of age, and 100 mL followed by at least 400 mL of water in children over 8 years of age.


1
R
L
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cm
(a)
(b)

37.16a,b

LONG-TERM COMPLICATIONS

Gastrointestinal problems after the newborn period are relatively common in children with CF. These include intussusception, appendiceal distension with inspissated material and appendicitis, rectal prolapse, and gallbladder disease. In the early 1990s, colonic strictures were reported by several centers in association with high pancreatic enzyme replacement. One of the most common gastrointestinal disorders in children and adolescents with CF is distal intestinal obstruction syndrome. The obturator obstruction often occurs after an intercurrent illness in which the child has a decreased oral intake and stops taking the pancreatic enzyme supplement.

Complicated meconium ileus

Neonates with complicated meconium ileus are managed in a similar manner to uncomplicated cases, although return of bowel function may be somewhat slower, particularly in cases of perforation. Some infants may require total parenteral nutrition if bowel function is slow to return, or if a proximal enterostomy does not provide an adequate absorptive surface to support the infant with enteral nutrition alone. Infants with both a proximal and distal stoma can often be managed with oral feeds combined with refeeding of the proximal ostomy effluent into the distal stoma. Enterostomy closure is generally performed 5–6 weeks after the initial procedure.

OUTCOME

The results of the medical and surgical management of 60 neonates with meconium ileus treated between 1972 and 1991 at the Riley Hospital for Children, Indianapolis, USA, have been reviewed. The study included 20 girls and 40 boys. A family history of CF was present in six neonates.

Twenty-five neonates had uncomplicated meconium ileus due to intraluminal obstruction of the terminal ileum with concretions of abnormal meconium. The treatment of these patients can be divided into two time periods, 1972–1980 and 1981–1991. Ten infants presented during the first time period, and only two of them were successfully cleared with a diatrizoate meglumine (Gastrografin) enema. The eight remaining infants underwent resection, operative irrigation, and enterostomy formation. A Bishop–Koop stoma was constructed in two infants, and six had a double-barrel (side-by-side) enterostomy. Of the 15 neonates treated during the later time period, eight (53%) were successfully cleared with a Gastrografin enema. The remaining seven infants required laparotomy. Seven were treated with enterotomy and intraoperative irrigation with saline or dilute contrast agent (Hypaque or Gastrografin) and one with irrigation and double-barrel enterostomy.

Thirty-five neonates presented with 56 complications of meconium ileus, including volvulus (22), atresia (20), perforation (6), and giant cystic meconium peritonitis (8). Clinical presentation in these neonates included abdominal distension, bilious vomiting, and failure to pass meconium; these symptoms were usually noted earlier than in uncomplicated cases. Neonates with perforation and giant cystic meconium peritonitis often had abdominal distension at the time of delivery. Three were diagnosed by prenatal ultrasound.

Operative management of patients with atresia, volvulus, and perforation included resection and anastomosis in 15 and enterostomy in 12. The eight patients with giant cystic meconium peritonitis underwent excision of the pseudocyst and enterostomy.

The diagnosis of CF was confirmed in all cases by sweat chloride test. Pancreatic enzyme therapy was instituted, along with a routine formula feed. Enterostomy closure was usually accomplished between 4 weeks and 3 months of age. All patients have been followed by the Indiana University Cystic Fibrosis Clinic at Riley Hospital for Children. Survival at 1 year was 92% (23/25) in patients with uncomplicated meconium ileus, and 89% (31/35) in complicated cases. The mortality in the uncomplicated cases was due to pulmonary problems and both occurred during the early time period. Deaths in the complicated cases were the result of sepsis (2), renal failure (1), and severe cholestatic jaundice progressing to liver failure (1). Since this report, uncomplicated cases have been managed exclusively at our institution with either enterotomy and irrigation, or irrigation through the appendix.

VIDEOS

OPS_8e_video_37.1 Meconium ileus: Initial evaluation.
OPS_8e_video_37.2 Meconium ileus: Operative procedures.

FURTHER READING

Burke MS, Ragi JM, Karamanoukian HL, et al. New strategies in nonoperative management of meconium ileus. *J Pediatr Surg* 2002; 37: 760–4.

Fitzgerald R, Colon K. Use of the appendix stump in the treatment of meconium ileus. *J Pediatr Surg* 1989; 24: 899–900.

O'Halloran SM, Gilbert J, McKendrick OM, et al. Gastrografin in acute meconium ileus equivalent. *Arch Dis Child* 1986; 61: 1128–30.

Rescorla FJ, Grosfeld JL. Contemporary management of meconium ileus. *World J Surg* 1993; 17: 318–25.

Sathe M, Houwen R. Meconium ileus in cystic fibrosis. *J Cyst Fibros* 2017; 16: S32–S39.

38

Vitellointestinal (omphalomesenteric) duct anomalies

SPENCER W. BEASLEY

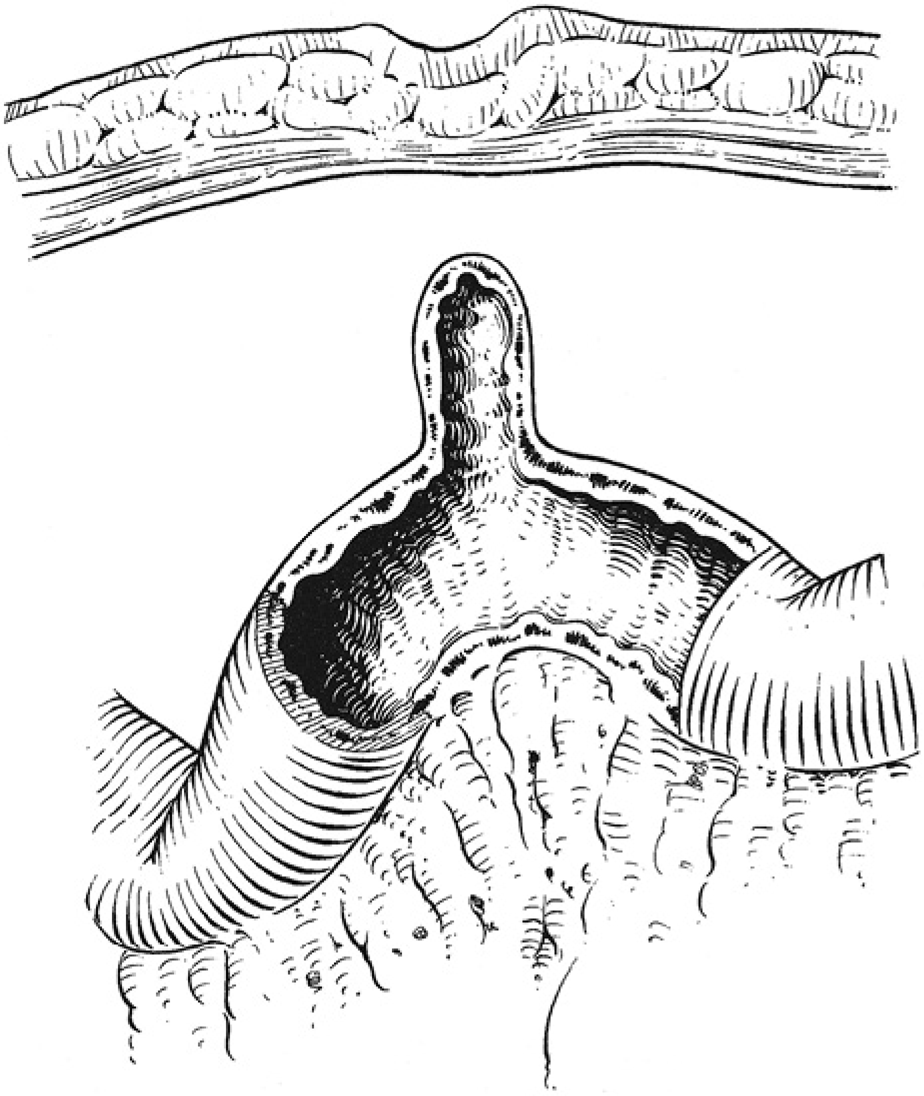
INTRODUCTION

The vitellointestinal (omphalomesenteric) duct is an embryonic communication between the yolk sac and the midgut. This communication normally disappears at about the sixth week of fetal life. Persistence of the duct between the intestinal tract and the umbilicus, or persistence of its embryonic blood supply, results in a variety of lesions that usually present in early infancy, but occasionally appear later in life.

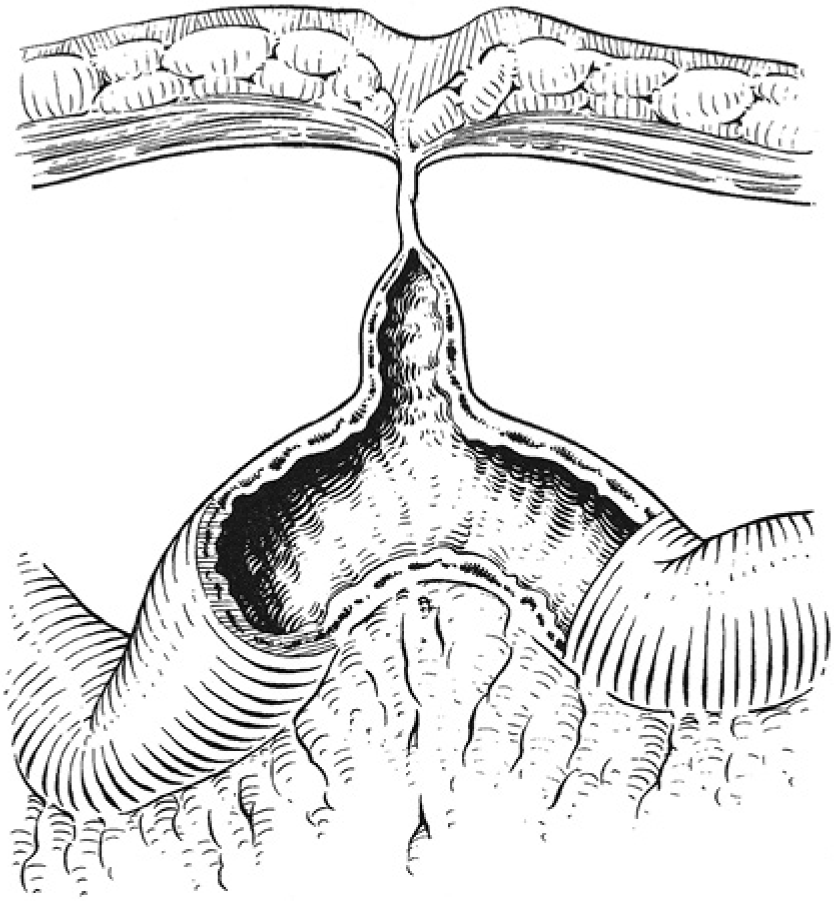
The structure is named for Johan Frederich Meckel (the Younger) (1781–1883). He was a noted German embryologist and published his observations on the diverticulum in 1809.

TYPES OF ANOMALY

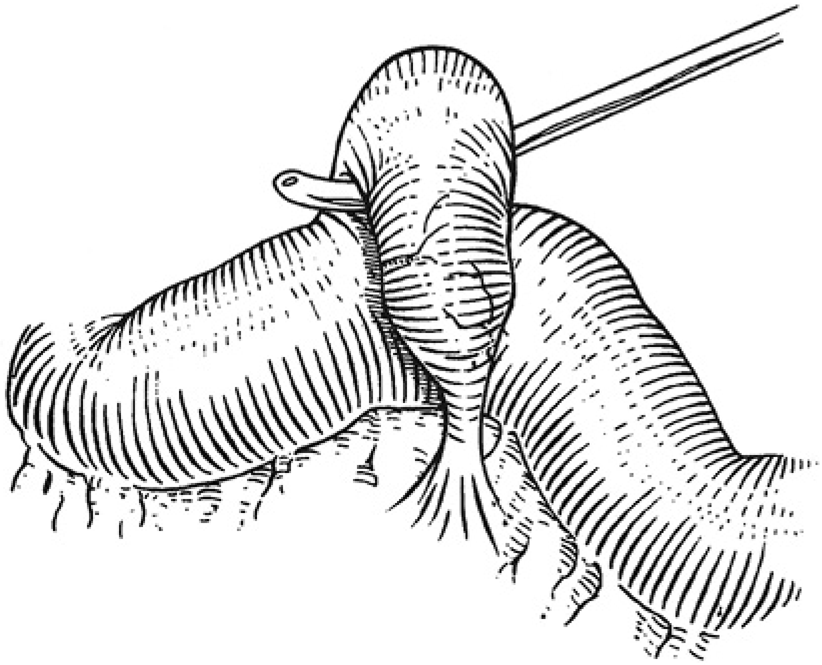
A Meckel's diverticulum represents persistency and patency of the inner intestinal component of the vitellointestinal tract (**Figure 38.1**). In a small proportion of patients there will be a fibrous band extending from the apex of the Meckel's diverticulum to the undersurface of the umbilicus (**Figure 38.2**), but more often there is a band representing the remnants of the vitelline vessels joining the Meckel's diverticulum to the mesentery of the small bowel (**Figure 38.3**). Meckel's diverticulum has diverse clinical presentations (**Table 38.1**), but frequently remains quiescent throughout life. The chances of an asymptomatic Meckel's diverticulum causing symptoms later in life are such that *en passant* removal of the structure when it is observed during operation for some other reason is arguably justified in children, but is probably not justified in adults in whom the chances of it becoming symptomatic are much less likely.



38.1



38.2



38.3

Table 38.1 Presentation of Meckel's diverticulum

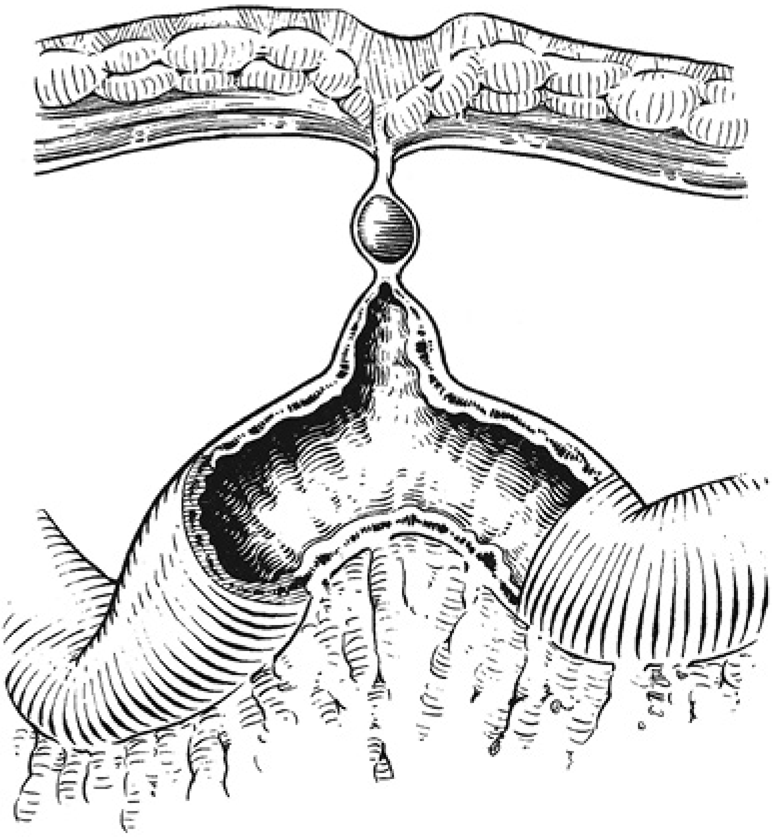
Melena and anemia	Ectopic gastric mucosa releases hydrochloric acid, which causes ulceration of adjacent ileum.
Abdominal pain (Meckel's diverticulitis)	Inflammation causes clinical features similar to those of acute suppurative appendicitis. Perforation of an adjacent ileal ulcer may also cause an inflammatory mass, pneumoperitoneum and peritonitis.
Intussusception	May invert and act as a lead point for an intussusception. This is responsible for about 2% of intussusceptions.
Meckel's band obstruction	A band extending from a Meckel's diverticulum to the root of the small bowel mesentery or the umbilicus may cause a loop of bowel to become entangled around it, producing a bowel obstruction.
Incidental findings	May be found during laparotomy for other conditions, e.g. appendicitis.

The typical appearance of a Meckel's diverticulum is shown in cross-section in Figure 38.1. Parts of the inner surface may contain ectopic pancreatic or gastric mucosa. Ectopic gastric mucosa produces hydrochloric acid, which can ulcerate adjacent non-gastric mucosa, either in the diverticulum itself or in adjacent ileum, and cause major bleeding. The child may present with “brick-red” rectal bleeding (often without much abdominal pain), and anemia.

The vitellointestinal band (Meckel’s band) is the remnant of the duct in which the lumen has been obliterated, but a fibrous cord or band persists. This runs from the deep surface of the umbilicus to the ileum or to a Meckel’s diverticulum. There is always a risk that a loop of bowel may become entangled around it, producing intestinal obstruction.

The Meckel’s diverticulum is often bound down by remnants of the vitelline vessels from its apex and is adherent to the mesentery of the ileum. (Division of the vessels at the apex of the peritoneal fold reveals its typical antimesenteric origin.) Loops of bowel can be trapped beneath this band, producing a closed loop obstruction.

This form of Meckel’s diverticulum has a cord containing a cystic remnant, which may slowly increase in size (**Figure 38.4**). The cyst is the result of partial obliteration of the duct. It may become infected and form an abscess; if it has a sinus, pus may discharge from the umbilicus.



38.4

BLEEDING MECKEL'S DIVERTICULUM

PRINCIPLES AND JUSTIFICATION

This is the most common clinical presentation and may result in rapid hemorrhage, which usually presents as relatively painless but profuse "brick-red" rectal bleeding. The resultant anemia may necessitate blood transfusion, but the bleeding usually stops spontaneously without the need for emergency surgery. The definitive investigation is surgery, but a technetium scan may confirm the presence of ectopic gastric mucosa.

INDICATIONS

Surgery (open or laparoscopic) is indicated where the clinical presentation of major and painless intestinal hemorrhage is consistent with a bleeding Meckel's diverticulum, irrespective of the result of the Technetium scan.

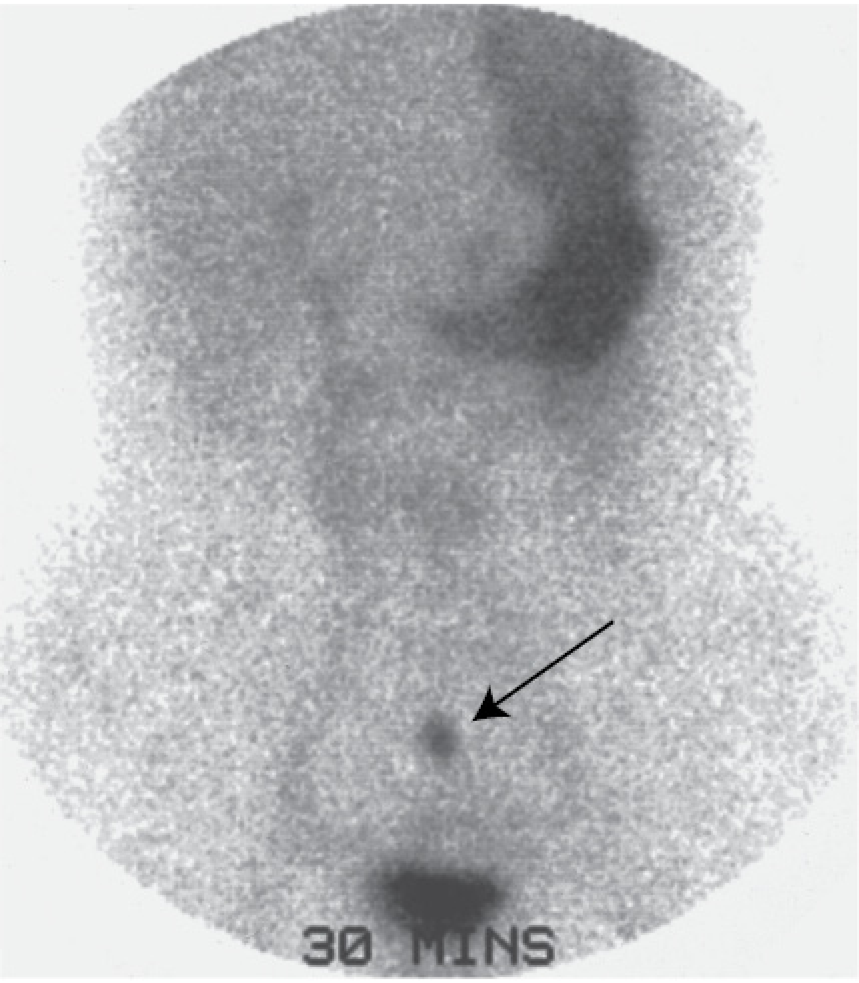
ANESTHESIA

The procedure is performed under general anesthesia. Blood transfusion is required very occasionally in the perioperative period, although the operative procedure itself is relatively bloodless. Perioperative antibiotics effective against bowel organisms are administered.

OPEN OPERATION

Incision

A right transverse, infraumbilical, muscle-splitting or muscle-cutting incision can be used (**Figure 38.5**). This can be extended medially by retracting the rectus toward the midline or even dividing the muscle if necessary. It can be excised easily through the standard incision used for an appendicectomy. Alternatively, laparoscopic-assisted Meckel's diverticulectomy through an umbilical incision can be used (see "Laparoscopic approach").



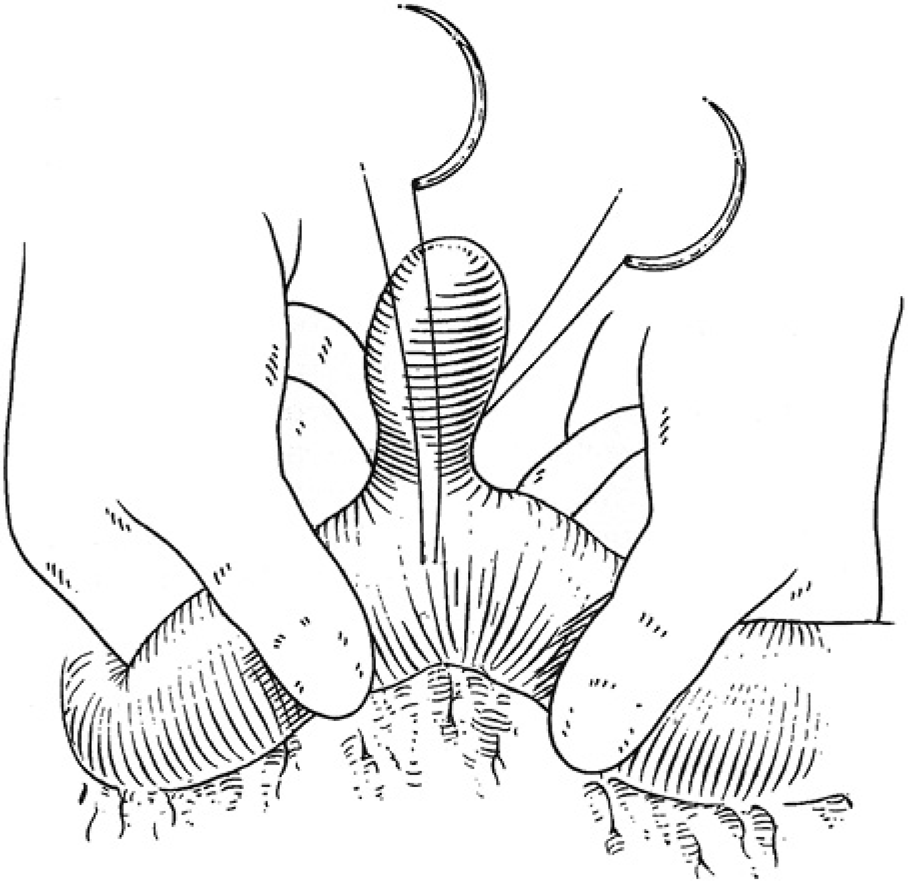
38.5

Identification of lesion

At laparotomy for bleeding, the Meckel's diverticulum will not usually be inflamed. A blue discoloration will be seen in the ileum and colon distal to the diverticulum if recent bleeding has occurred.

Control of ileum and its contents

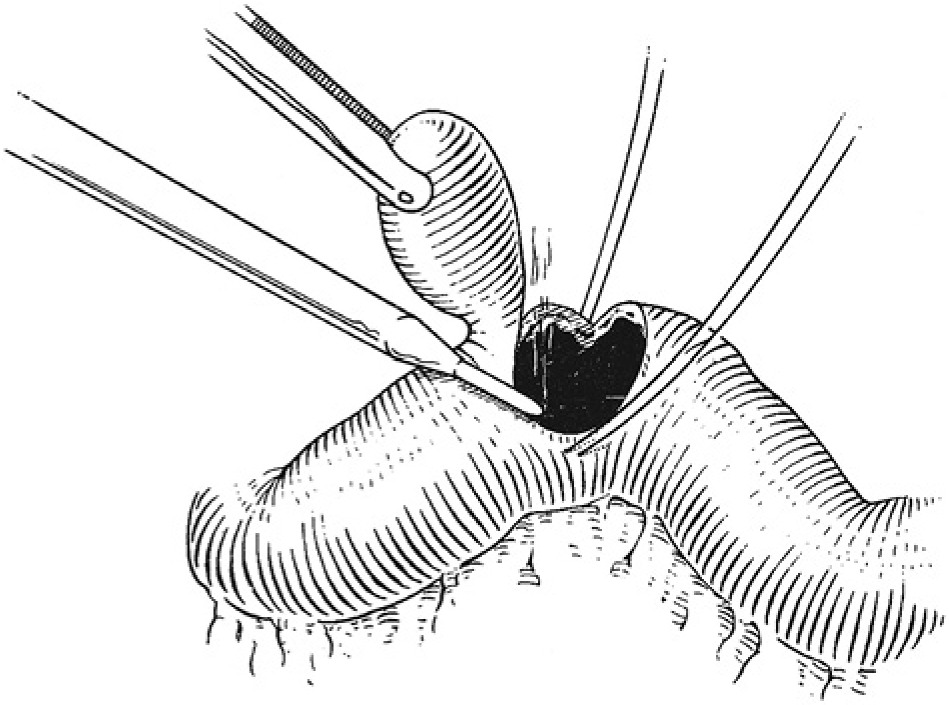
Compression of the ileum with the fingers or a non-crushing bowel clamp (fingers are preferred because they are less traumatic to the bowel) reduces the amount of bleeding and soiling that occurs when the ileum is opened and the diverticulum is excised (**Figure 38.6**). Packs are placed on either side of the loop of ileum containing the Meckel's diverticulum. Suction is kept nearby to reduce accidental spillage of liquid ileal contents when the ileum is opened. Stay sutures (3/0) are placed on the ileum on either side of the diverticulum.



38.6

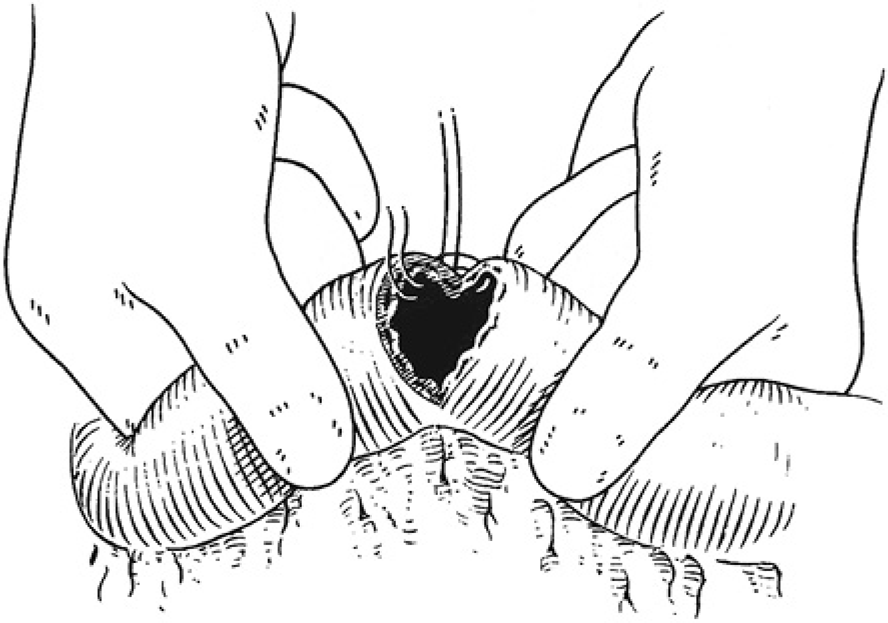
Excision of diverticulum

A longitudinal or oblique elliptical incision is made in the ileum near the base of the diverticulum using scissors or diathermy (**Figure 38.7**). It is essential that the entire diverticulum is removed, because a remnant of acid-secreting mucosa left at the base of the diverticulum could continue to cause ulceration and bleeding of the adjacent ileum.



38.7

The stay sutures are then held apart to transform the longitudinal or oblique elliptical incision into a transverse one (**Figure 38.8**). This allows closure of the wound without narrowing the lumen of the ileum.



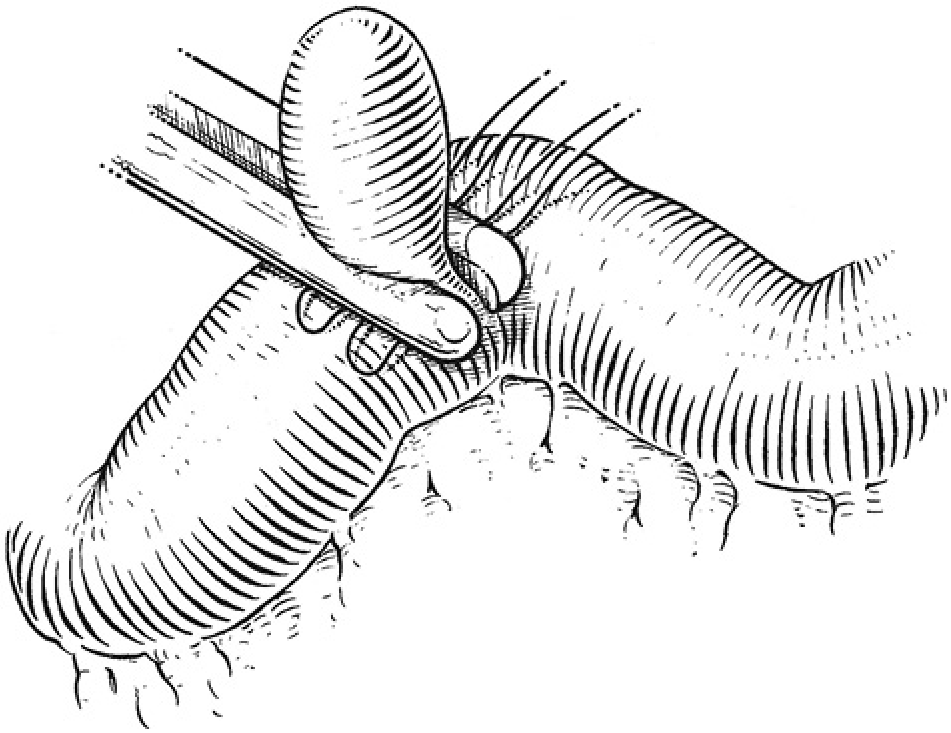
38.8

Closure of ileum

A 4/0 absorbable, continuous or interrupted, all-layers suture is used to close the bowel. A second seromuscular continuous layer is employed by some surgeons.

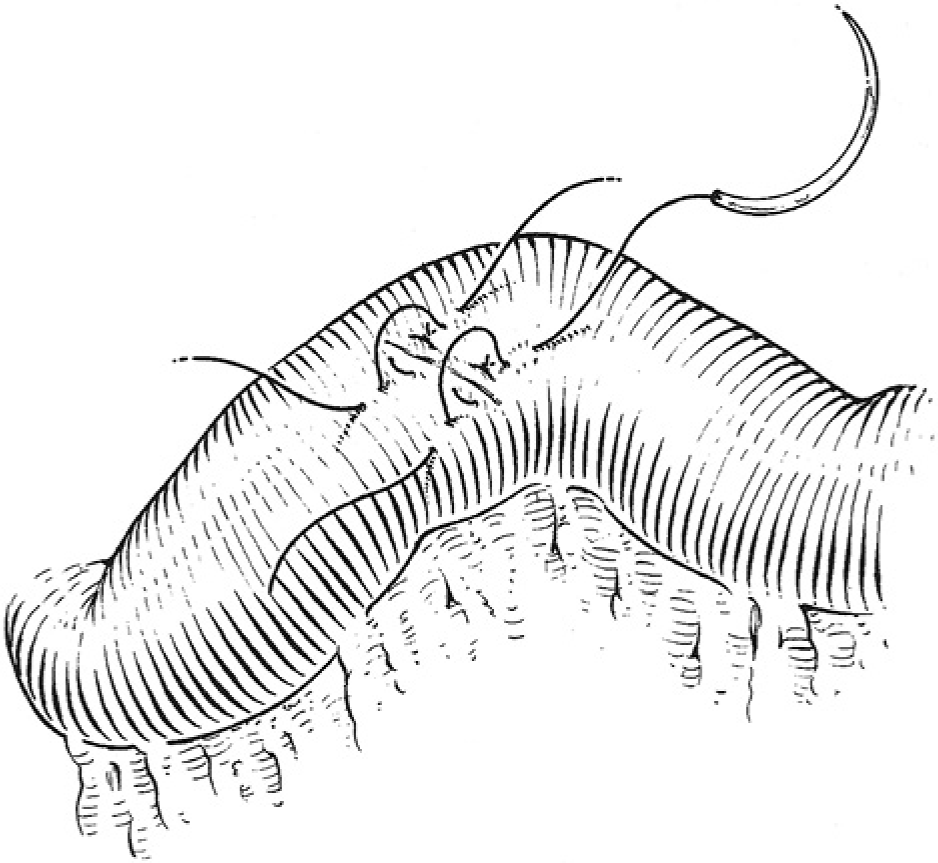
Alternative method for narrow-necked diverticulum

A curved artery forceps or crushing clamp is placed across the base of the diverticulum at 45 degrees or more to the long axis of the ileum (**Figure 38.9**). This avoids the narrowing that might be caused by an incision closed longitudinally. Mattress sutures of 3/0–4/0 absorbable material are inserted under the clamp and tied. The diverticulum is cut away at the distal border of the clamp.



38.9

The clamp is removed and the line of section is buried by a second layer of sutures (**Figure 38.10**).



38.10

The wound is closed in layers in the standard fashion.

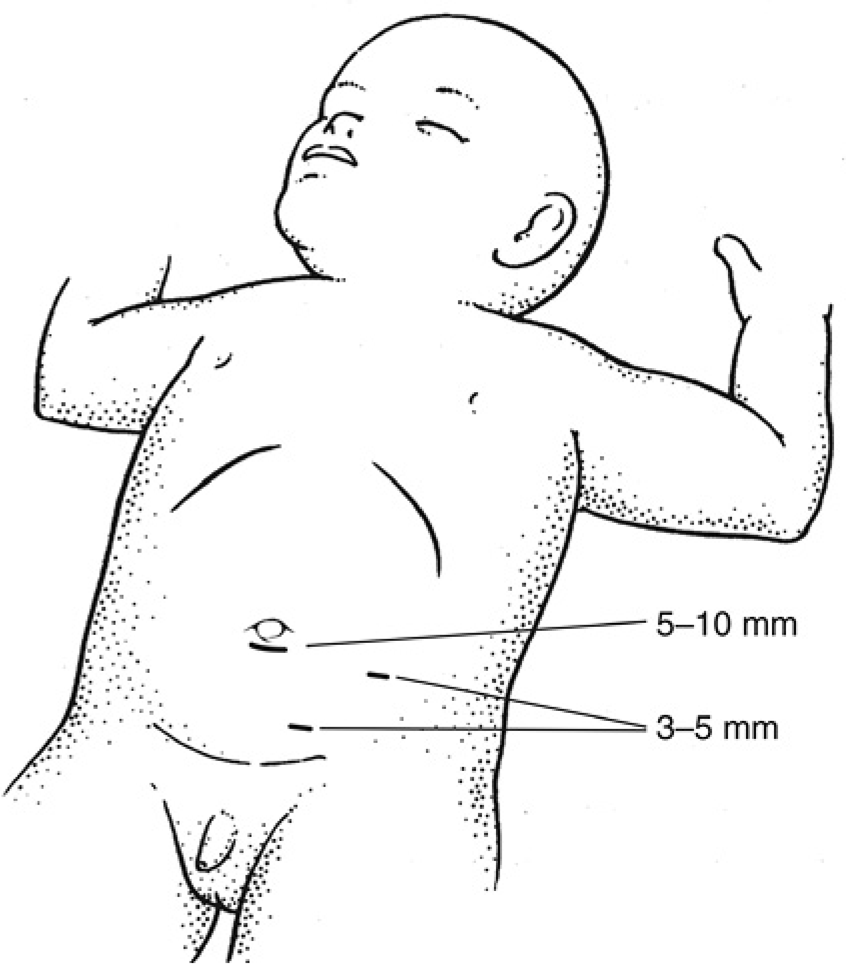
POSTOPERATIVE CARE

Oral fluids can be commenced on about the second post-operative day. Most children can be discharged on the second or third day after operation.

LAPAROSCOPIC APPROACH

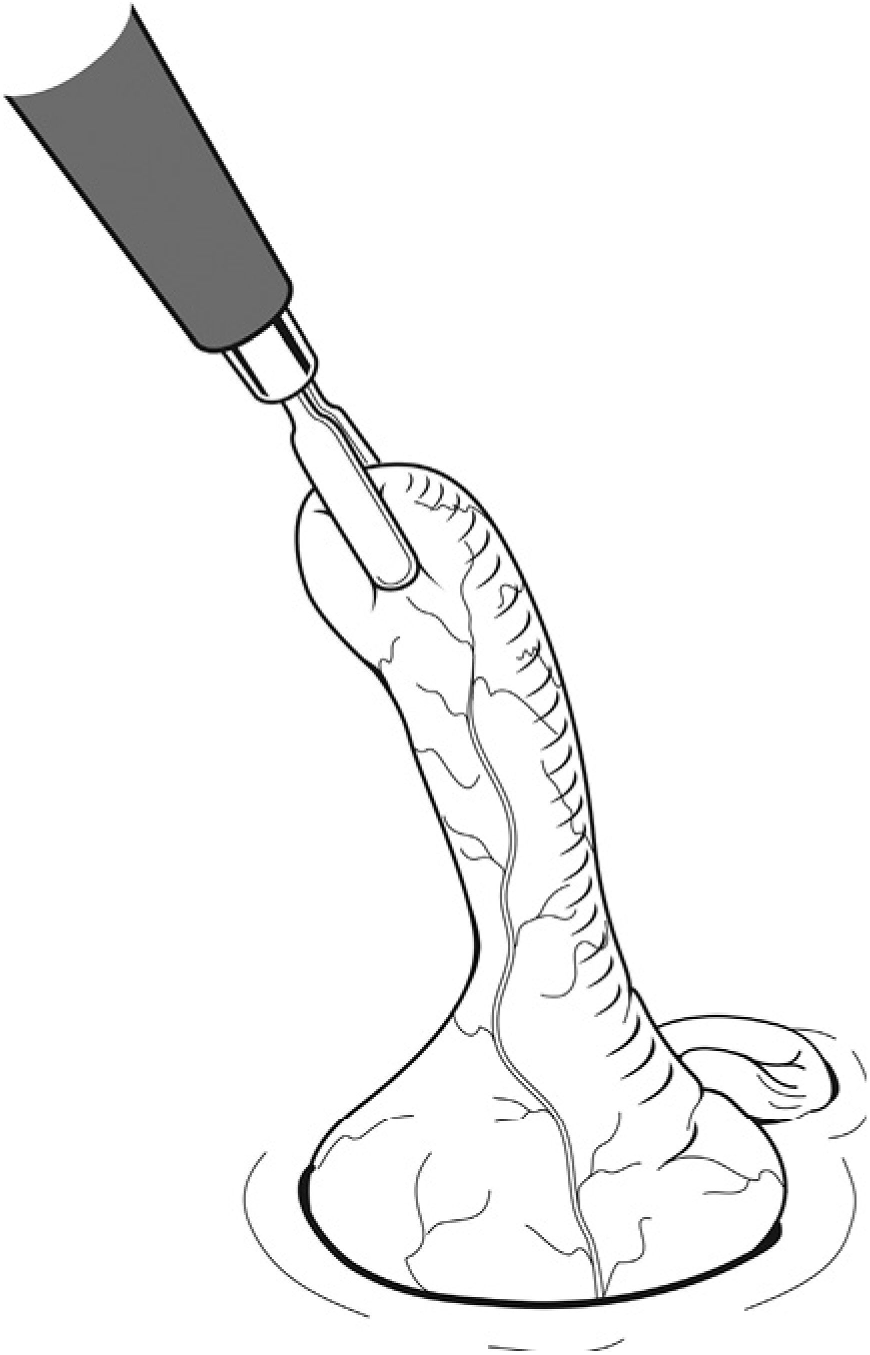
This is now the preferred approach of many surgeons, and offers the advantage of being able to confirm the presence of a Meckel's diverticulum using a minimally invasive technique where there has been diagnostic uncertainty preoperatively.

Under general anesthesia and infiltration of local anesthetic, an infraumbilical incision is made. The linea alba is incised and the peritoneum opened under direct vision. A 5 mm or 10 mm 30-degree telescope is introduced through a 5 mm or 10 mm umbilical port and secured to the rectus fascia. An initial inspection of the peritoneal cavity is performed. One or two 3 mm or 5 mm working ports can be introduced on the left side of the abdomen, the size and location being determined by the age of the child (**Figure 38.11**).



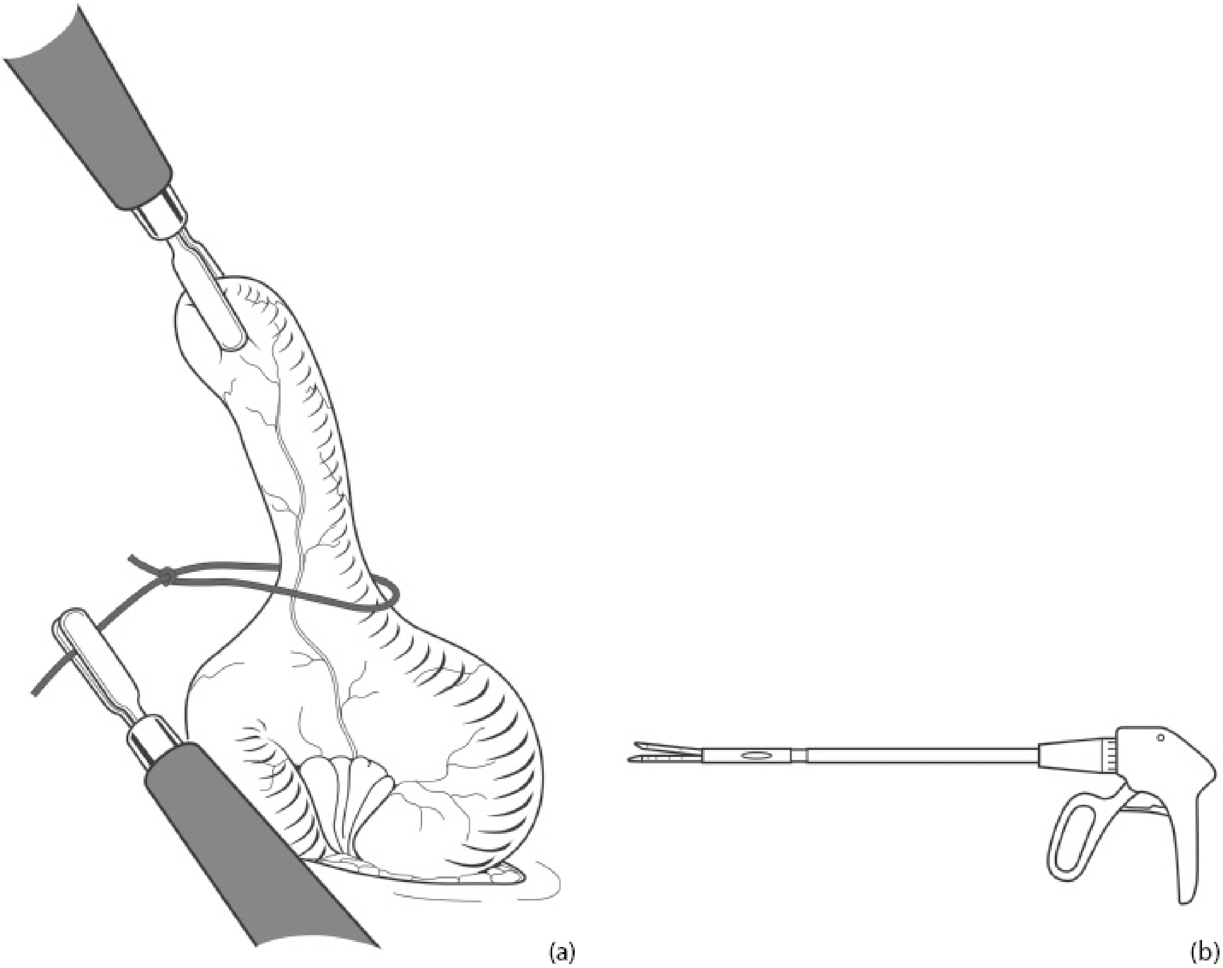
38.11

Once located, the Meckel's diverticulum can be delivered through the umbilical port opening (sometimes requiring enlargement) and is removed outside the abdomen using the same technique as described above (**Figure 38.12**).



38.12

Alternatively, particularly where the diverticulum is quite narrow at its base, it can be dissected from the ileum laparoscopically: The vitelline vessels are either divided by electrocautery or included in two ENDOLOOPS□ placed around the base of the Meckel's diverticulum (**Figure 38.13a**). Care is taken both to ensure the whole diverticulum is removed (to avoid leaving ectopic gastric mucosa behind) and to ensure that the caliber of the lumen of the ileum is not compromised. The Meckel's diverticulum is divided between the ENDOLOOPS□ and removed through the umbilical port. An alternative and equally effective method involves resection using an Endo GIA□ stapler (**Figure 38.13b**).



38.13a,b

MECKEL'S DIVERTICULITIS

PRINCIPLES AND JUSTIFICATION

Meckel's diverticulitis is an unusual presentation in children. When it does occur, the child is assessed clinically as having acute appendicitis, but at laparotomy the appendix is found to be normal and there is an inflammatory process involving a Meckel's diverticulum. In all operations for suspected appendicitis in which the appendix is found to be normal at laparotomy or on laparoscopy, the distal 100 cm of ileum should be inspected to exclude inflammation of the Meckel's diverticulum as being responsible for the symptoms. The inflamed Meckel's diverticulum is often palpable in the abdomen on opening the peritoneum. An inflammatory mass around a Meckel's diverticulum may result from perforation of an ulcer in the ileum adjacent to the base of the diverticulum. Perforation may also cause pneumoperitoneum and peritonitis.

OPERATION

Indication and exposure

When the diagnosis is known or suspected before the operation, a small, transverse, right infraumbilical incision can be used. If the diagnosis is made during operation at the time of exploration for suspected appendicitis, the inflamed Meckel's diverticulum and adjacent ileum can be delivered easily through the appendicectomy wound and the diverticulum excised outside the abdomen. If a laparoscopic approach to appendicectomy has been performed, the Meckel's diverticulum can be removed laparoscopically.

Extensive inflammatory mass

If there is severe or long-standing inflammation of the diverticulum causing edema and involving the surrounding ileum, or if the base of the Meckel's diverticulum is very broad, it is appropriate to excise it with a small sleeve of ileum and perform an end-to-end small bowel anastomosis along conventional lines.

SMALL INTESTINAL OBSTRUCTION

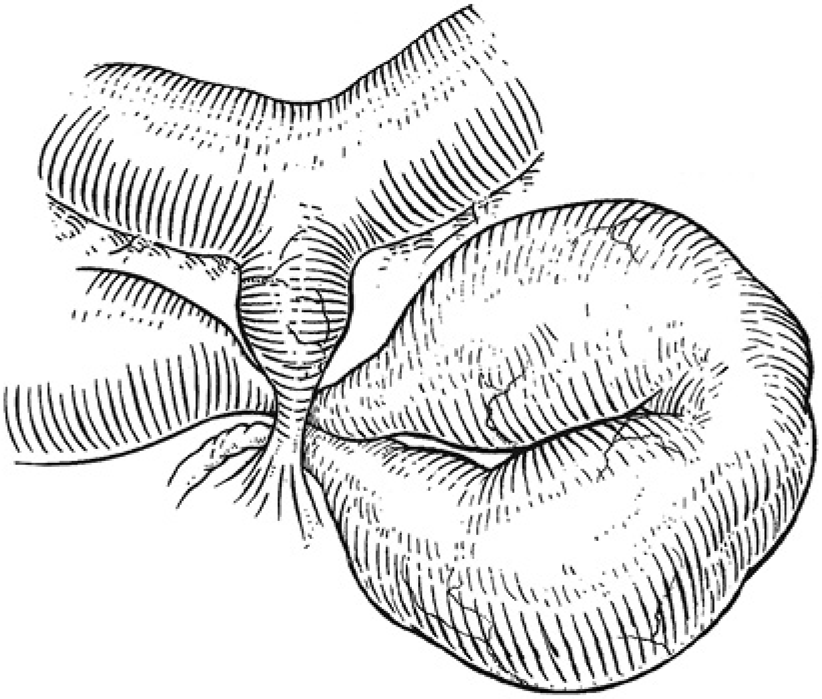
INDICATIONS

These children present with a distal small bowel obstruction, the exact cause of which usually cannot be determined clinically. Closed loop obstruction is common. A laparoscopic approach can be used by the experienced laparoscopic surgeon, but the distended loops of small bowel may compromise visualization of the cause of the obstruction.

OPERATION

Abdominal access (either open or laparoscopic) is achieved as described above.

The band running from the Meckel's diverticulum is identified and divided, releasing the entrapped loops of bowel (**Figure 38.14**). Most commonly, the band runs to the root of the small bowel mesentery, but may be attached to the undersurface of the umbilicus.



38.14

Occasionally, the small bowel trapped by the band in a closed loop obstruction is found to be gangrenous and will need to be removed. The ends of viable bowel are reconnected with a standard end-to-end anastomosis.

INTUSSUSCEPTION OF MECKEL'S DIVERTICULUM

Meckel's diverticulum is the most common cause of intussusception in which a pathologic lesion at the lead point is identified. It may occur at any age during childhood, but is most common in the first two years of life.

INDICATIONS

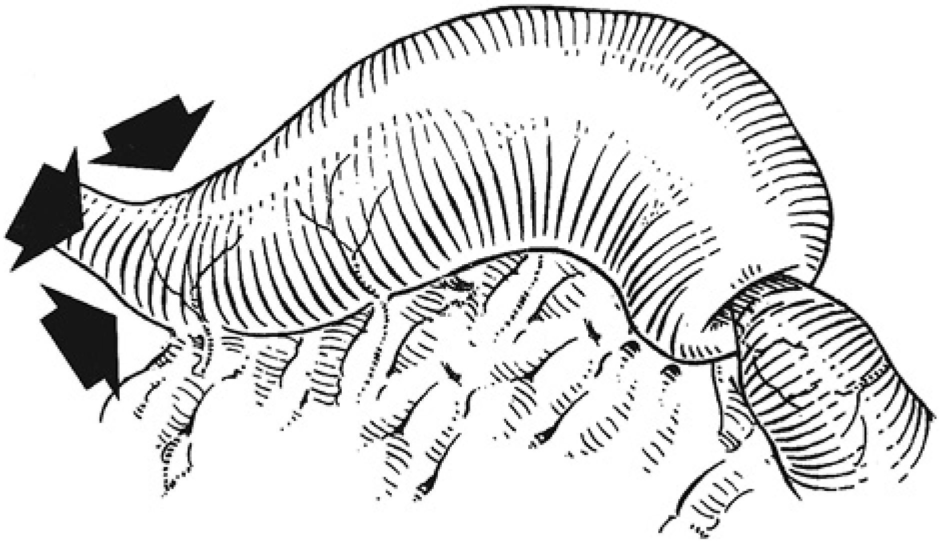
Peritonitis or other clinical evidence of the presence of ischemic bowel is an absolute indication for surgery in patients with intussusception. Otherwise, the child is treated initially by gas (or barium) enema. Where the intussusception is due to a Meckel's diverticulum, however, enema reduction is unlikely to be successful, and surgery is indicated.

OPERATION

Surgical approach

Abdominal access is achieved as described above.

The intussusception is located and an attempt is made to reduce it manually by gentle compression of the colon in a proximal direction at the level of the lead point (indicated by arrows in **Figure 38.15**). Much of the intussusception can be reduced, but when there is a Meckel's diverticulum at the lead point, the final portion often cannot be reduced and must be resected in continuity with the diverticulum.



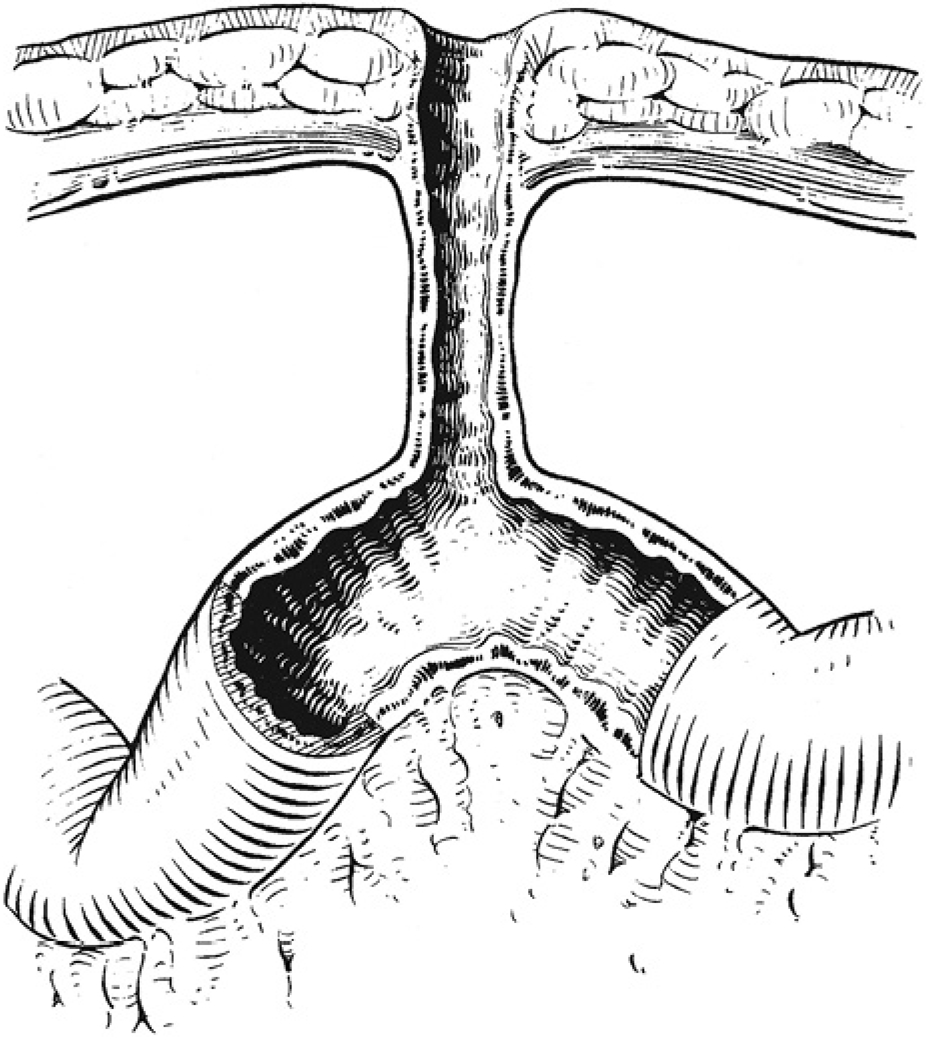
38.15

The laparoscopic approach relies more on gently easing the intussusceptum out of the intussuscipiens proximally using a rocking motion. When the need for resection becomes evident either because the intussusception cannot be reduced or because a Meckel diverticulum has been identified, the lesion can be delivered through an expanded umbilicus for extracorporeal resection. This may require the introduction of a 5 mm camera through one of the working ports to guide manipulation of the lesion as it is delivered through the umbilicus.

COMPLETE PERSISTENCE OF THE VITELLOINTESTINAL DUCT (FISTULA)

PRINCIPLES AND JUSTIFICATION

When the entire vitellointestinal (omphalomesenteric) duct persists and remains patent, there is an open communication between the ileum and the umbilicus (**Figure 38.16**). This allows intermittent discharge of the contents of the ileum (gas and ileal fluid) and causes periumbilical excoriation. The tract may be of sufficient caliber to allow prolapse of the ileum as a “pair of horns.”



38.16

DIAGNOSIS

Escape of air and feces through an opening in the umbilicus is pathognomonic of a patent vitellointestinal tract. When there is doubt about the nature of the discharge from the umbilicus (often because it is intermittent), a sinogram will usually demonstrate direct communication with the ileum.

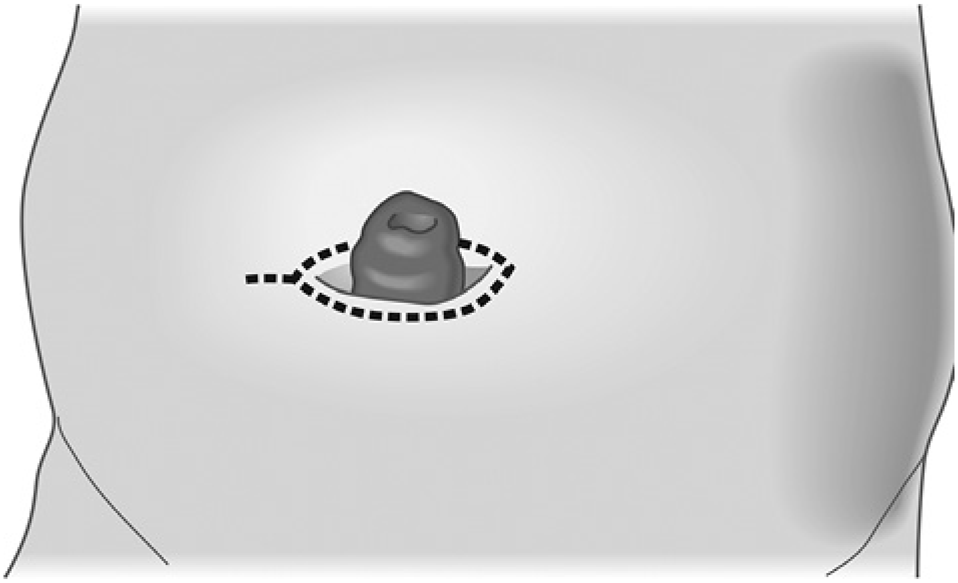
INDICATIONS

Surgery is indicated in all cases where a patent vitellointestinal tract has been shown.

OPERATION

Incision

An incision is made in the umbilicus at the mucocutaneous junction, circumcising the external opening of the patent vitellointestinal tract (**Figure 38.17**). The tract is mobilized by separating it from the tough fascia of the linea alba surrounding it, and the peritoneum is opened. Sometimes, to improve exposure, the incision may need to be extended lateral to the umbilicus by dividing the medial 1–2 cm of rectus abdominis muscle, including the anterior rectus sheath, medial fibers of the rectus abdominis muscle, and the posterior rectus sheath.



38.17

Dissection of tract

Dissection of the vitellointestinal tract is continued into the peritoneal cavity, where its communication with the ileum is identified (**Figure 38.18**).

38.18

Division at junction with ileum

Two stay sutures are placed on the ileum and the vitellointestinal tract is divided at this point. The communication with the ileum may be narrow, resembling a fibrous cord, or broad, as in many patients with a Meckel's diverticulum. The ileum is closed in one or two layers with 3/0 or 4/0 absorbable sutures.

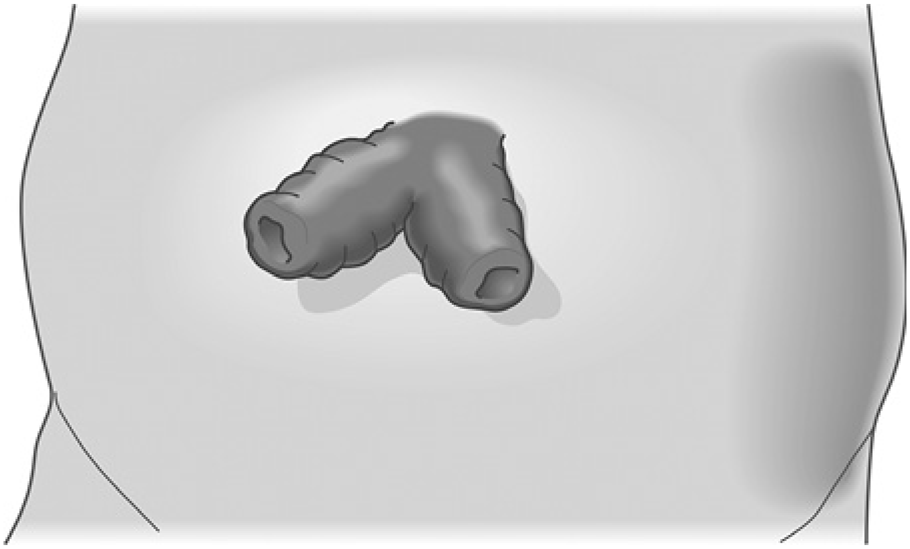
Reconstruction of umbilicus

The defect in the umbilical ring is repaired in the same way as for an umbilical hernia.

PROLAPSE OF PATENT VITELLOINTESTINAL TRACT

PRINCIPLES AND JUSTIFICATION

When the channel is a short and broad vitellointestinal tract, the ileum may intussuscept through it onto the surface of the umbilicus, producing a double-horned "Y-shaped" segment of bowel, inside out, with a lumen evident on each horn (**Figure 38.19**).



38.19

OPERATION

As long as the bowel is not necrotic (necrosis is rare), it should be reduced manually, after which the vitellointestinal tract can be excised as a semi-elective procedure. In extremely rare situations, there is a single-horned prolapse of the vitellointestinal tract. This is seen in the neonatal period and suggests that there is an atresia of one horn, which will necessitate excision of the vitellointestinal tract and end-to-end ileoileostomy to reconstitute gastrointestinal continuity. This is usually an isolated lesion, not associated with other abnormalities.

ECTOPIC MUCOSA

PRINCIPLES AND JUSTIFICATION

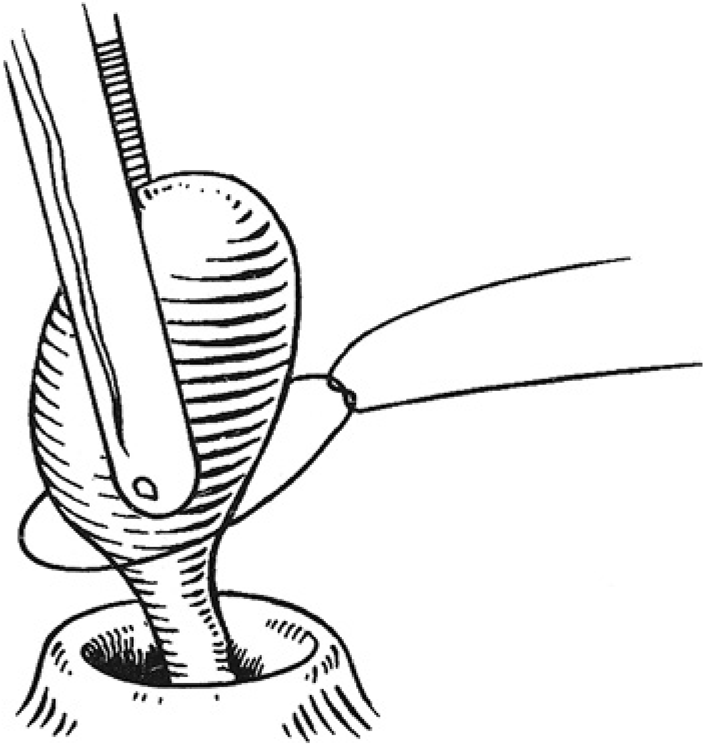
Discharge of mucus from the umbilicus may be caused by a small sequestrated nodule of ectopic alimentary mucosa (representing persistence of the external part of the vitellointestinal tract). It appears as a shiny, spherical, deep-red nodule in the depths of the umbilical cicatrix, but may be pedunculated. Crusts may form on the cicatrix and on the surrounding skin. It should be distinguished clinically from the more common umbilical granuloma, which presents as a small mass of heaped pink or grayish granulation tissue producing a chronic seropurulent discharge; the granuloma can be treated with topical application of silver nitrate.

CONTRAINDICATIONS

If a sinus opening is evident, it is likely that all or part of the vitellointestinal tract is present and patent. Likewise, discharge of air or fecal fluid suggests complete patency of the vitellointestinal tract (vitellointestinal fistula). In this situation, surgical excision of the whole tract will be required.

OPERATION

Pedunculated ectopic mucosa can be ligated within the umbilical ring (**Figure 38.20**); alternatively, a wider based lesion may be excised by diathermy dissection across its base. Suture is required only occasionally.



38.20

FURTHER READING

Alemayehu H, Stringel G, Lo IJ et al., Laparoscopy and complicated Meckel diverticulum in children. *JSLS* 2014; 18(3): e2014.00015.

Campbell J, Beasley SW, McMullin N, Hutson JM. Clinical diagnosis of umbilical swellings and discharges in children. *Med J Aust* 1986; 145: 450–3.

Loh DL, Munro FD. The role of laparoscopy in the management of lower gastrointestinal bleeding. *Pediatr Surg Int* 2003; 19: 266–7.

Potts SR. Meckel's diverticulum. In: Najmaldin A, Rothenberg S, Crabbe D, Beasley S (eds). *Operative Endoscopy and Endoscopic Surgery in Infants and Children*. London: Hodder Arnold; 2005: pp. 273–7.

Sanders LE. Laparoscopic treatment of Meckel's diverticulum: Obstruction and bleeding managed with minimal morbidity. *Surg Endosc* 1995; 9: 724–7.

Saw RS. Appendix and Meckel diverticulum. In: Oldham KT, Colombani PM, Foglia RP (eds). *Surgery of Infants and Children: Scientific Principles and Practice*. Philadelphia, PA: Lippincott-Raven; 1997: pp. 1224–8.

Shalaby RY, Soliman SM, Fawy M, Samaha A. Laparoscopic management of Meckel's diverticulum in children. *J Pediatr Surg* 2005; 40: 562–7.

Valla JS, Steyaert H, Leculée R et al. Meckel's diverticulum and laparoscopy of children. What's new? *Eur J Pediatr Surg* 1998; 8: 26–8.

39

Duplications of the alimentary tract

STIG SØMME AND JACOB C. LANGER

HISTORY

Duplications of the alimentary tract have also been termed giant diverticula, enterogenous cysts, ileum duplex, and inclusion cysts. Calder described the first case in 1733. Ladd introduced the term "duplications of the alimentary tract" in 1937. He also listed a set of three criteria for the diagnosis:

- There will be a well-developed coat of smooth muscle.
- Epithelial lining will represent some part of the alimentary tract.
- There will be an intimate attachment to some part of the alimentary tract.

PRINCIPLES AND JUSTIFICATION

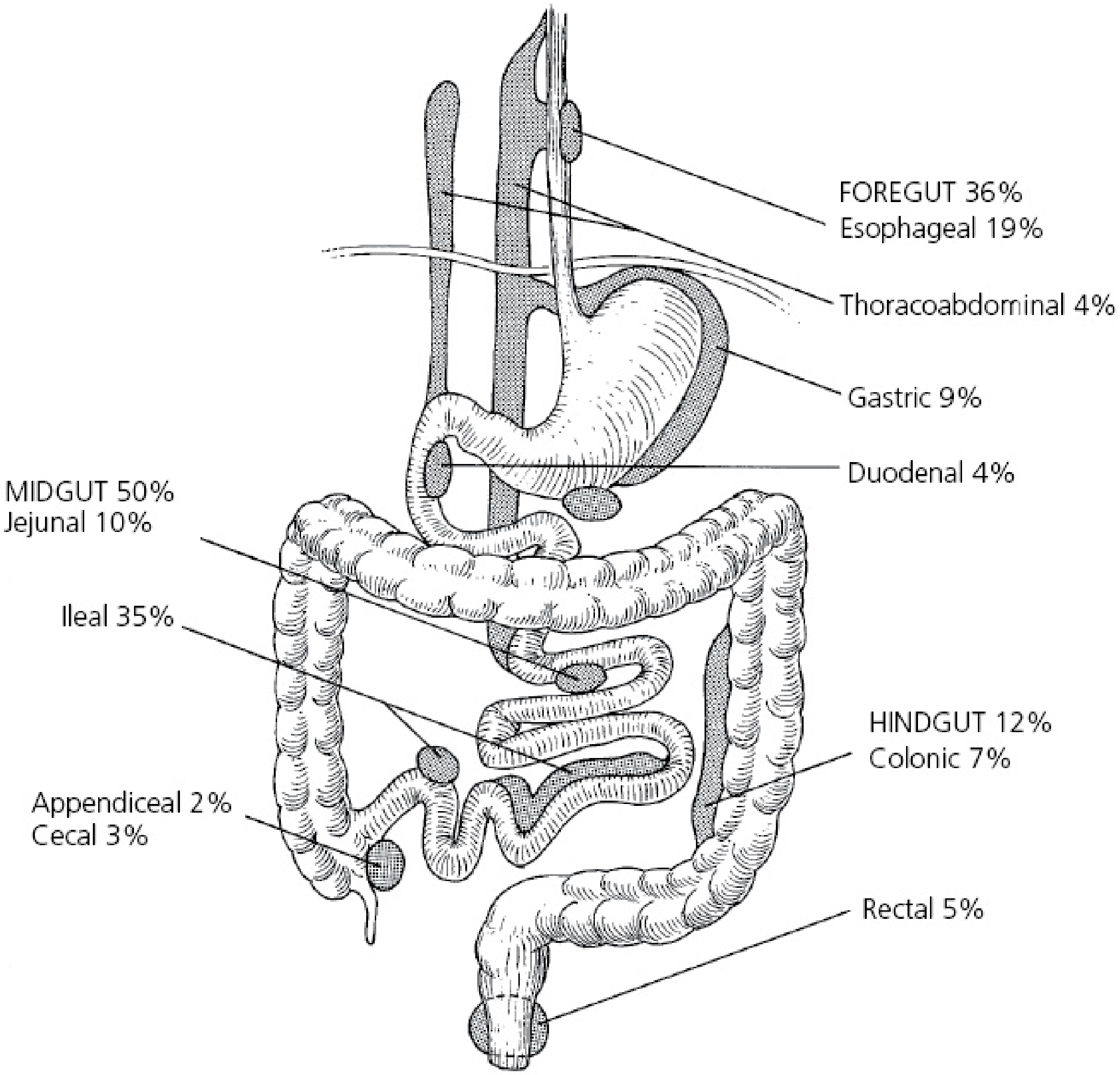
Alimentary tract duplications are congenital malformations that may be found anywhere from mouth to anus. Duplications are found in about 1 in 4500 autopsies. The most common duplication is cystic and located on the mesenteric aspect of the small or large intestine. Multiple duplications are seen in about 10% of patients. As many as 30% of patients with thoracic or thoracoabdominal duplications have additional duplications below the diaphragm. Duplications are lined by alimentary tract mucosa and usually share a common smooth muscle wall and blood supply with the adjacent gut. Ectopic acid-producing gastric mucosa may be present in approximately

20% of duplications, and can cause peptic ulceration, bleeding, and perforation. Most duplications cause symptoms in infancy or early childhood and most are diagnosed by the age of 2 years, but some present as an incidental finding in patients with other conditions or may be identified on routine prenatal ultrasound; about 50% are diagnosed before 5 months of age. There is an overall equal sex distribution. In adults, duplications can rarely present with adenocarcinoma.

The pathogenesis for the development of duplications is unknown, but cystic duplications are thought to develop secondary to a split notochord mechanism and tubular duplications secondary to partial twinning. Cystic duplications may be associated with spinal cord and vertebral anomalies, and tubular duplications may be associated with urinary tract, spine, and central nervous system anomalies. Foregut duplications probably occur during the normal division of the foregut into respiratory and esophageal structures, and may therefore contain elements of respiratory tract, such as cartilage and bronchial epithelium (bronchogenic cyst), elements of esophagus such as smooth or striated muscle and squamous epithelium (esophageal duplication), or elements of both (foregut duplication).

DISTRIBUTION AND VARIETY

The distribution of 495 alimentary tract duplications in 455 patients shows that the most common location is in the midgut, and most of these duplications are located in the ileum (**Figure 39.1**).


FOREGUT 36%
Esophageal 19%
Thoracoabdominal 4%
Gastric 9%
Duodenal 4%
MIDGUT 50%
Jejunal 10%
Ileal 35%
Appendiceal 2%
Cecal 3%
HINDGUT 12%
Colonic 7%
Rectal 5%

39.1

PRESENTATION

Patients may present with a mass or with symptoms related to the location and size of the cyst. Intestinal duplications can serve as a lead point for intussusception and can cause a localized volvulus of the small bowel. Ectopic acid-producing gastric mucosa can cause peptic ulceration of the normal intestinal mucosa, with resultant painless gastrointestinal bleeding or perforation. Duplication cysts may also cause intestinal obstruction due to mass effect, particularly those in the small bowel, which are located on the mesenteric surface between the leaves of the mesentery. Foregut duplication cysts may present with obstructive symptoms involving the esophagus, trachea, or both. Some duplication cysts are diagnosed on routine prenatal ultrasound. The differential diagnosis in these cases includes ovarian cyst, mesenteric or omental cyst, lymphatic malformation, choledochal cyst, and, less commonly, cysts of solid organs such as the kidney, spleen, liver, or pancreas. A pediatric surgeon should counsel the parents prenatally. Postnatally, the infant should be evaluated by a pediatric surgeon and should have a repeat ultrasound. If the child is asymptomatic and the cyst is still present and less than 4 cm in diameter, it is reasonable to manage it expectantly with repeated ultrasounds and a technetium scan to rule out ectopic gastric mucosa. Ovarian cysts will usually resolve over time, whereas duplication cysts will not. Symptomatic cysts should be resected expeditiously.

TREATMENT

Because alimentary tract duplications may be complicated by infection, bleeding, intestinal obstruction or volvulus, and may lead to the development of malignancy, many surgeons believe that they should be treated by early complete excision when possible. Sometimes complete excision is not achievable, for example in cases of long segmental tubular duplications or complex duodenal or retroperitoneal duplications. Development of adenocarcinoma of the mucosal lining is rare, with only a few cases described in the literature. It is therefore acceptable to manage asymptomatic duplications that are not amenable to resection expectantly. Unresectable symptomatic duplications should be managed by partial resection in order to deal with the particular symptom. With close monitoring it is safe to wait until an infant is 3–12 months old before resecting asymptomatic duplication cysts. At 3–12 months infants are larger, tolerate the surgery well and are potentially less affected by anesthesia and risks of infection.

PREOPERATIVE ASSESSMENT AND PREPARATION

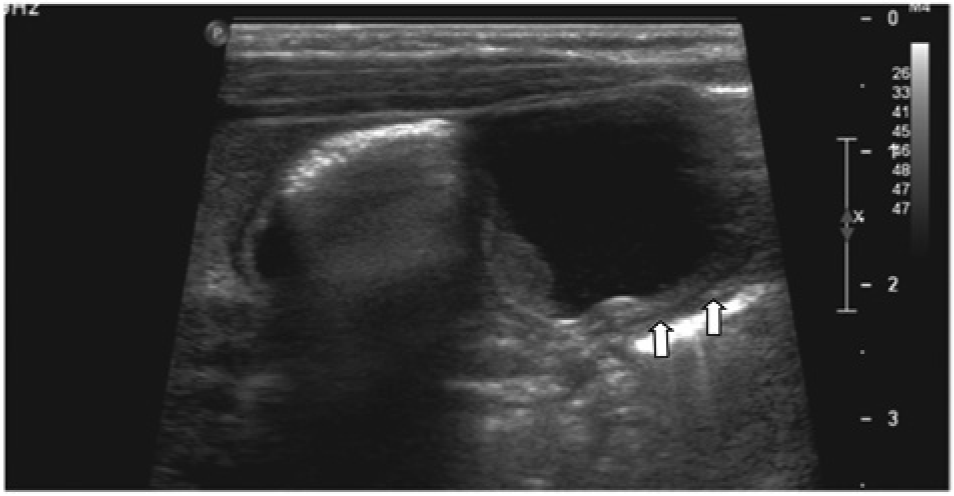
Associated anomalies

Associated anomalies are rare, but can be predicted based on the location of the duplication. Foregut duplications may be associated with vertebral anomalies. Midgut duplications may have associated malrotation or atresia of the bowel. In patients with hindgut lesions, genitourinary duplications, anorectal malformations, and bladder exstrophy are sometimes seen. Associated anomalies are often identified clinically or with imaging. When multiple anomalies are found in the same patient, it is important that the management of the patient be planned carefully, since a multidisciplinary approach involving various specialists may be necessary.

Imaging diagnostics

ULTRASOUND

Ultrasound will determine whether a mass felt on physical examination is cystic or solid and its relationship to the intestine. Often a characteristic outer hypoechoic rim and an inner echogenic rim can be seen, a sign that is sometimes referred to as a "bowel signature" (**Figure 39.2**).



39.2

ABDOMINAL RADIOGRAPHS

Radiographs of the abdomen may reveal a mass effect in the case of large abdominal cysts. Chest X-ray may show a mass, usually in the middle or posterior mediastinum, and associated vertebral anomalies.

COMPUTED TOMOGRAPHY AND MAGNETIC RESONANCE IMAGING

Computed tomography (CT) and/or magnetic resonance imaging (MRI) may be helpful in cases where ultrasound is unclear or cannot be done for technical reasons. These modalities are particularly useful for the investigation and delineation of foregut duplications, duodenal duplications, and rectal lesions. Chest CT with contrast is useful for the visualization of foregut duplications, and provides important information for the preoperative planning of the procedure. MRI may be helpful in identifying and characterizing involvement of the spinal cord in cases of neurenteric cysts.

TECHNETIUM SCANNING

Technetium scanning is used to identify ectopic gastric mucosa in the cyst, which is present in about 20% of cases. Routine use of this technique is not necessary, but it can be very useful in the investigation of the child with painless rectal bleeding, for which the differential diagnosis includes both Meckel's diverticulum and intestinal duplication. It is also useful in the investigation of the child with an asymptomatic prenatally diagnosed duplication, in whom demonstration of ectopic gastric mucosa may push the surgeon to recommend excision rather than observation.

ESOPHAGOGASTRODUODENOSCOPY

Esophagogastroduodenoscopy may be helpful to identify ulcers or strictures, and to better define the anatomy prior to operative excision of duplication cysts in the upper gastrointestinal tract.

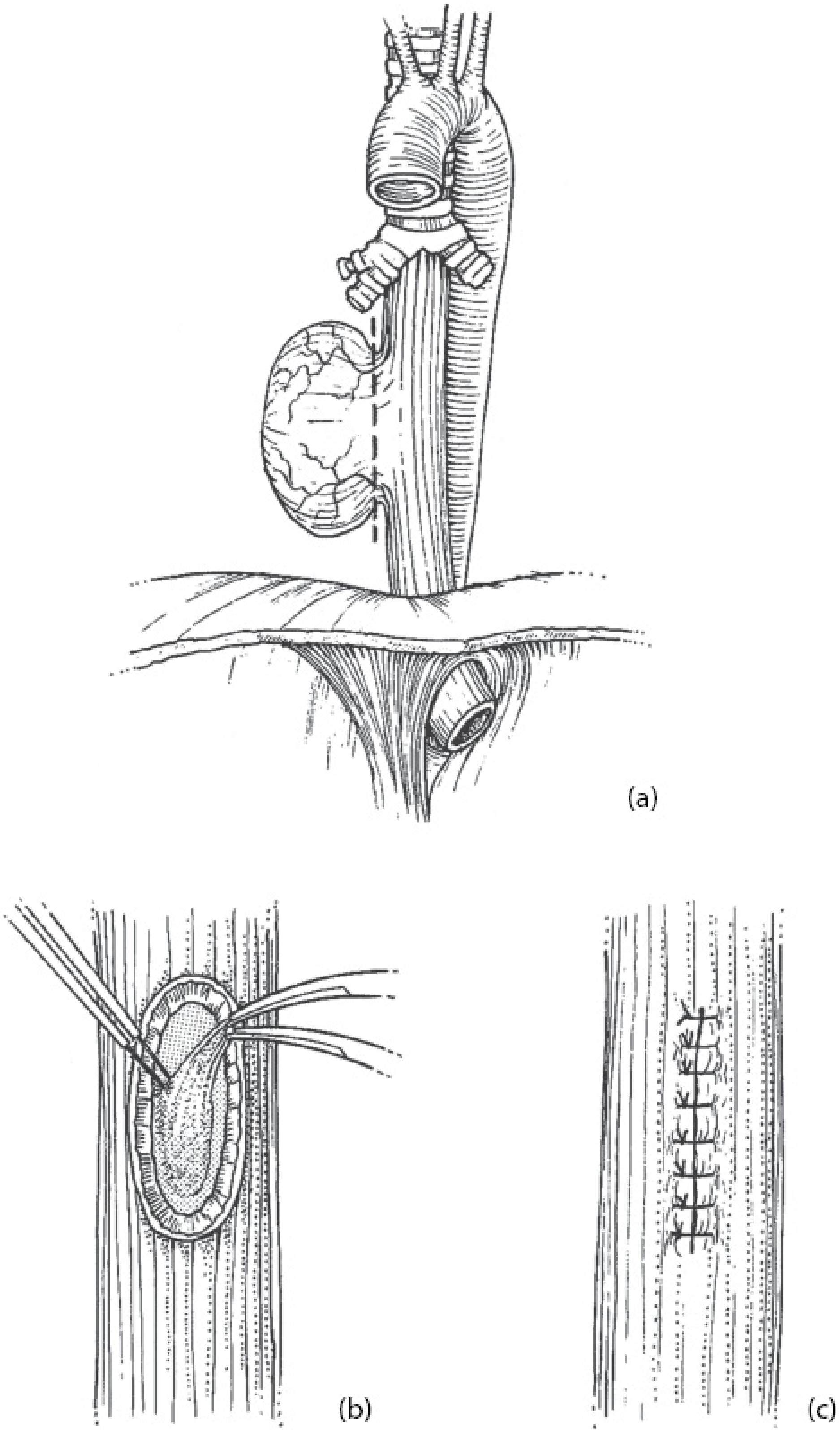
CONTRAST RADIOGRAPHY

Upper and lower intestinal contrast radiography may be helpful for elucidating the anatomy. Barium swallow will demonstrate a typical anterior indentation of the esophagus in cases of foregut duplication cysts.

OPERATIONS

Esophageal duplications

Esophageal duplication cysts and foregut duplication cysts are usually intramural, non-communicating, cystic lesions. They are more commonly located to the right side of the esophagus (**Figure 39.3a**). Those in the proximal esophagus are often situated between the esophagus and the trachea. A posterolateral left or right thoracotomy provides excellent exposure for most esophageal duplications, although many surgeons are now using a thoracoscopic approach (see "Minimally invasive surgery for duplications"). Proximal foregut duplications may be removed more easily through a supraclavicular cervical incision. The phrenic and vagus nerves and the thoracic duct should be carefully identified to avoid injury. Esophageal duplication cysts are excised by dissecting the duplication off the esophageal wall at the base; it is important to remove residual mucosa from the wall of the esophagus (**Figure 39.3b and c**) to avoid recurrence. This can be accomplished by stripping or using fulguration with electrocautery. If the esophagus is inadvertently entered, it can be repaired primarily and as needed, a pleural flap can be used to buttress the repair. If an esophageal stricture or ulcer was noted on preoperative esophagoscopy or swallow study, it may be necessary to perform a segmental resection of the esophagus.

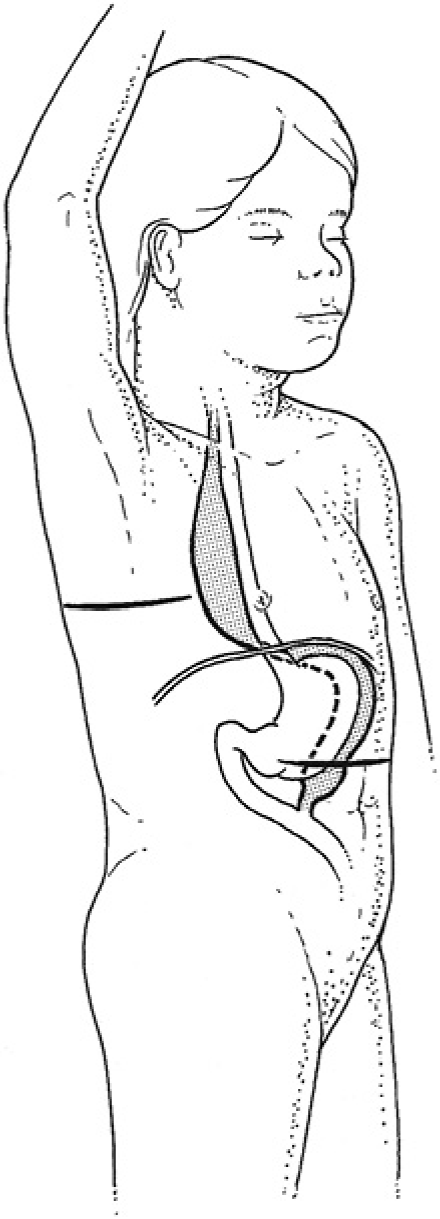


39.3a–c

Attempts should be taken to reapproximate the muscularis over the excised area, since weakness of the esophageal wall with subsequent diverticulum formation can occur if the submucosa is not contained.

Thoracoabdominal duplications

Thoracoabdominal duplications are commonly located to the right of the esophagus in the posterior mediastinum. They may communicate with the stomach or small intestine through the diaphragm, and may also communicate with the spinal canal (neurenteric cysts). These lesions are best approached with a combination of thoracotomy/thoracoscopy and laparotomy/laparoscopy, or through a thoracoabdominal incision (**Figure 39.4**).

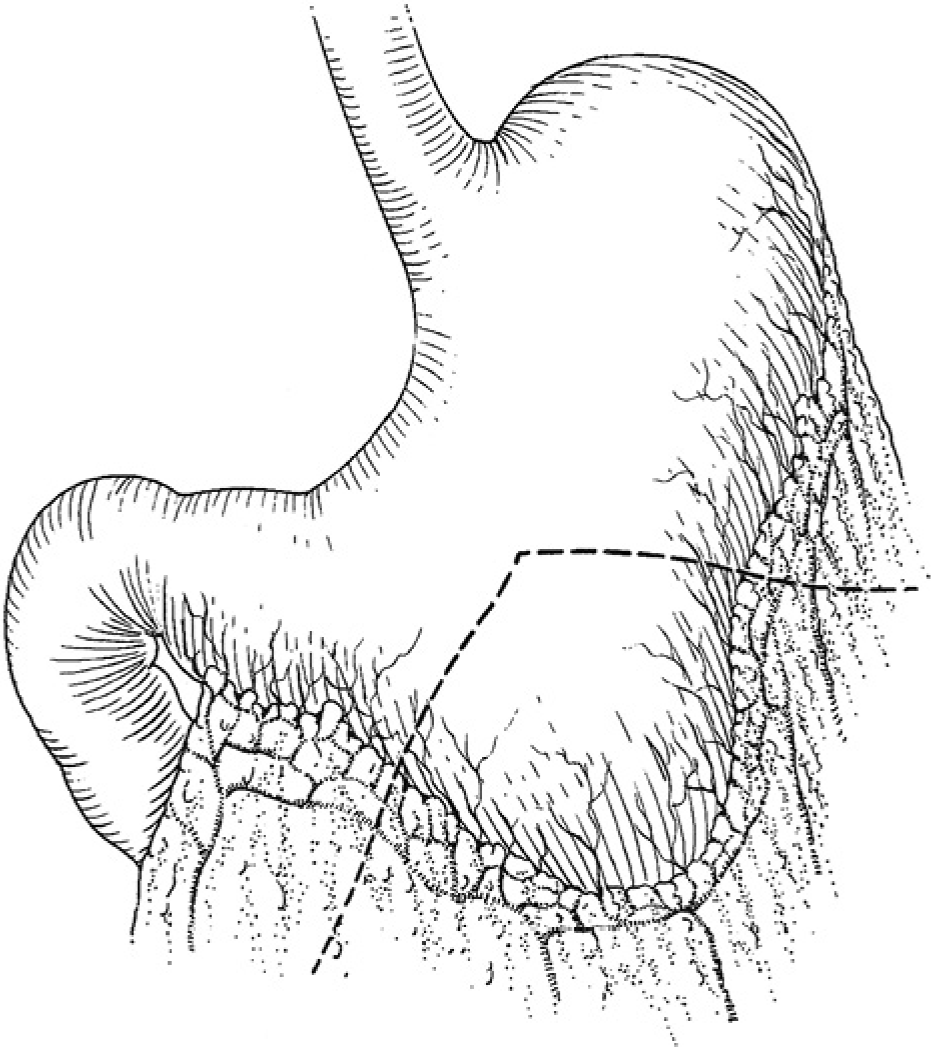


39.4

It is recommended to start with a posterolateral thoracotomy or thoracoscopy and to excise the intrathoracic component first. This involves dissecting the duplication off any attachments in the chest. Sometimes it is attached to the vertebrae and must be carefully dissected off the bone. If an intraspinal component is present and a laminectomy is necessary for complete removal, a neurosurgeon should be involved. After the duplication is free of any attachments in the chest, it is traced distally to the diaphragm, where it is divided between ligatures. Next, the resection is completed through a laparotomy or laparoscopy. The duplication is easier to identify below the diaphragm if the abdomen is opened prior to the completion of the dissection in the chest. That way, pulling on the duplication will help identify its continuation in the abdomen. The diaphragmatic defect can usually be closed primarily. The abdominal part of the duplication is often a tubular lesion communicating with the small intestine, although it sometimes ends blindly along the greater curve of the stomach. Occasionally, thoracoabdominal duplications present with hemoptysis as peptic ulceration leads to erosion into the lung.

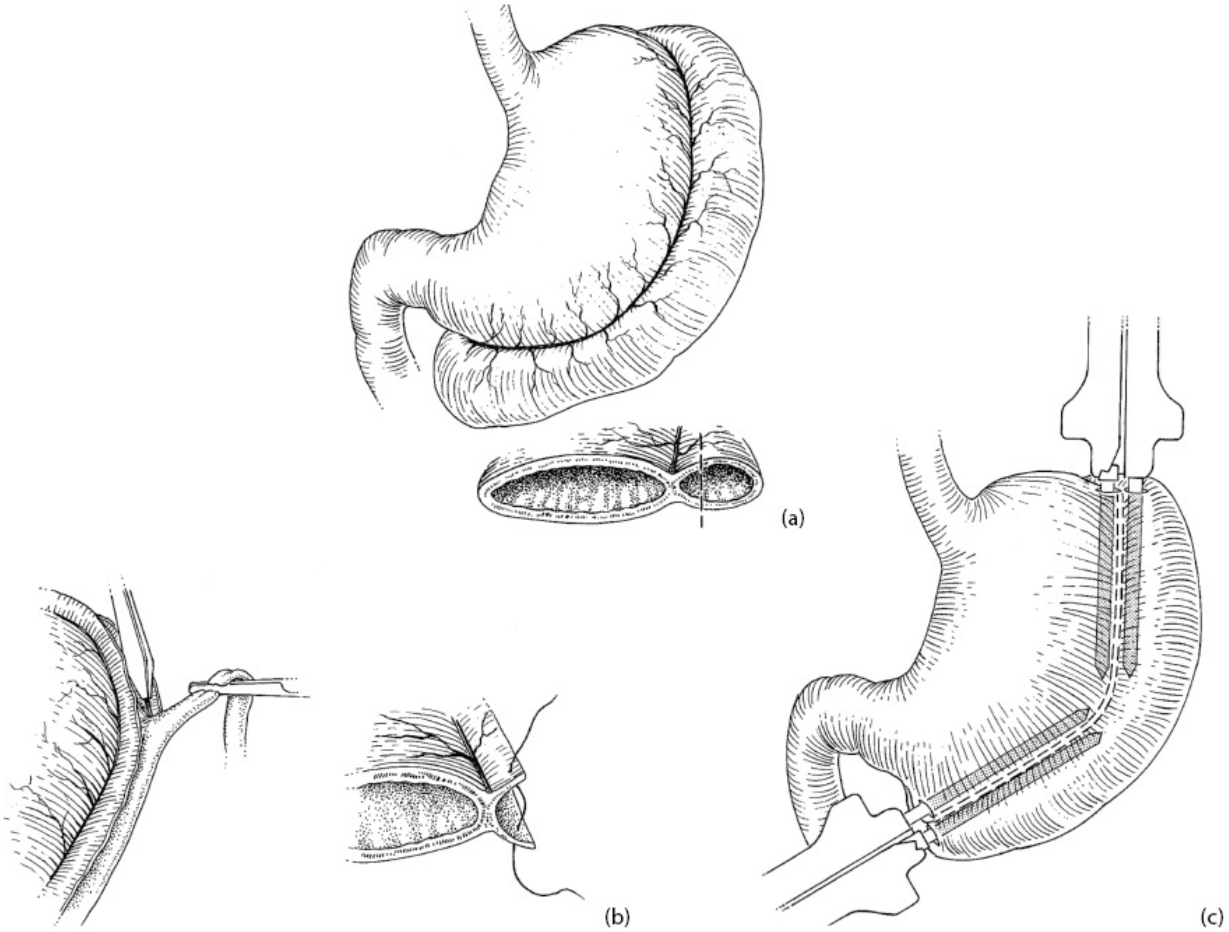
Gastric duplications

Duplications of the greater curvature can be treated by complete resection (**Figure 39.5**). As with esophageal duplications, most gastric duplications are contained within the stomach wall and can be resected without entering the gastric lumen. Laparoscopic or laparoscopic-assisted resection has been described.



39.5

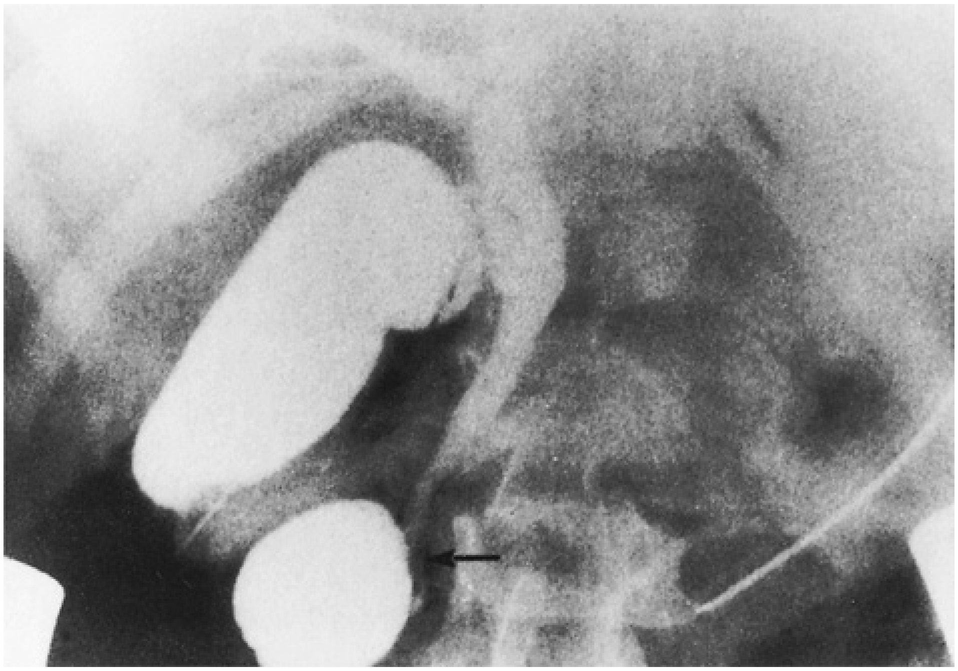
If the duplication cyst involves an extensive part of the stomach, partial resection and stripping of the residual mucosal lining is an alternative (**Figure 39.6a–c**).



39.6a–c

Duodenal duplications

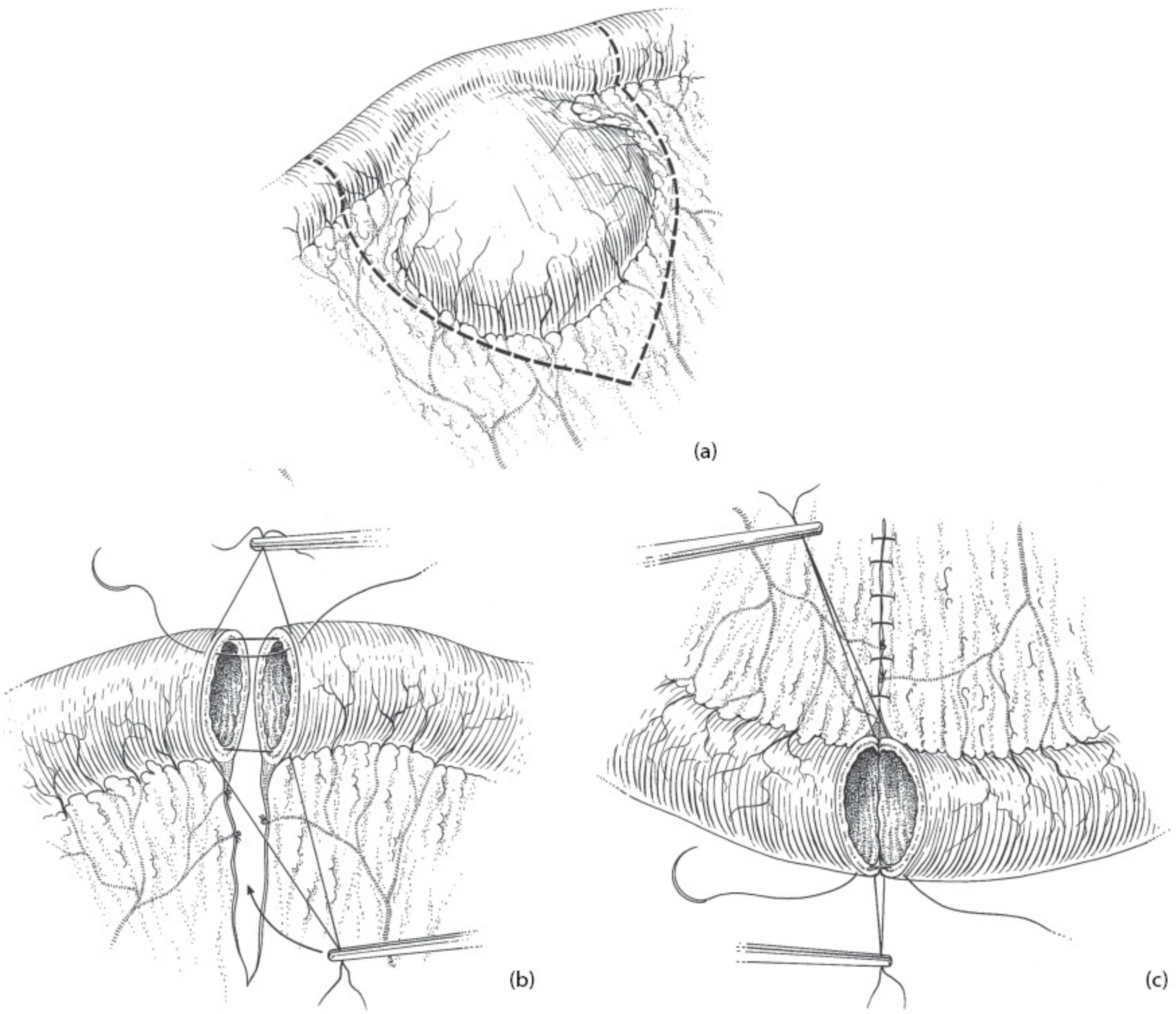
Duplications of the duodenum may present with vomiting and abdominal pain, but due to the close proximity to the biliary tree and pancreas, symptoms and findings related to these organs can be part of the initial presentation. Duodenal duplication cysts of the first, third, and fourth portion can usually be excised using a technique similar to that described for esophageal and gastric duplications. Duodenal duplications attached to the second portion of the duodenum should undergo preoperative and/or intraoperative radiographic visualization of the bile ducts and pancreatic ducts (**Figure 39.7**). If the biliary or pancreatic ducts are involved, complete excision will be difficult. Partial excision and mucosal stripping are an acceptable alternative in these cases. In some cases, a small bowel Roux-en-Y loop can be brought up and a cystenterostomy created, similar to what has been described for the treatment of pancreatic pseudocysts.



39.7

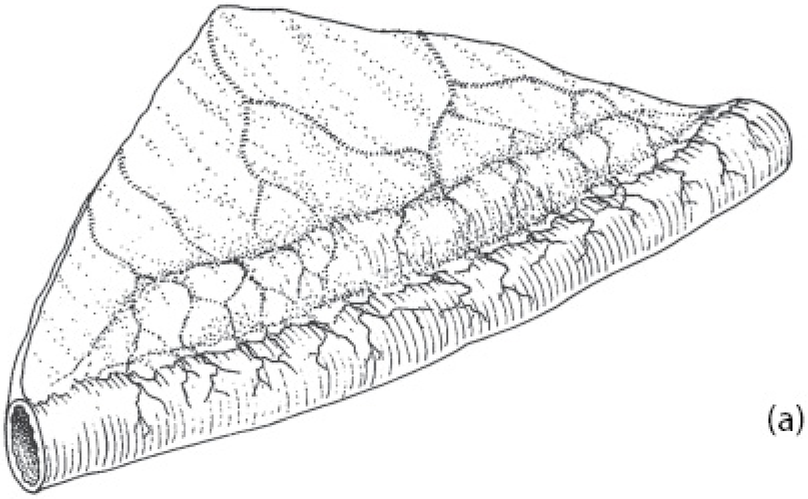
Small intestinal duplications

Cystic lesions of the jejunum or ileum are the most common duplications and are usually excised without difficulty. Because cyst and normal bowel often share a common muscular wall and blood supply, they are usually resected as one specimen (**Figure 39.8a–c**). This procedure can often be performed using laparoscopic techniques.

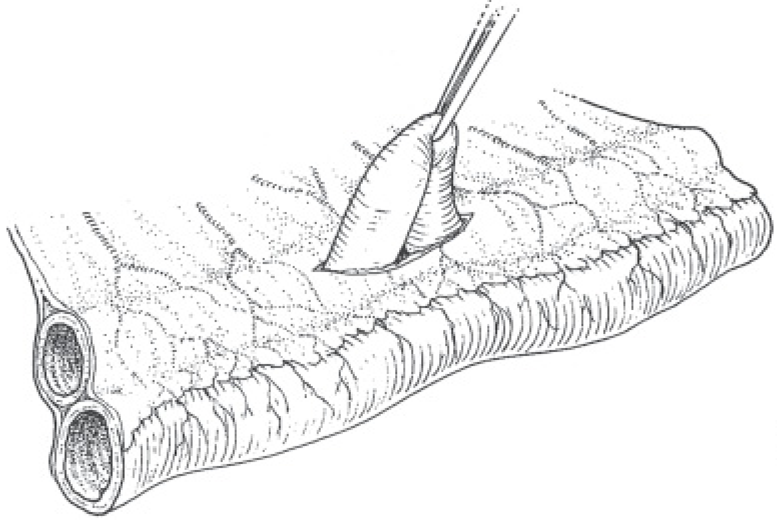


39.8a–c

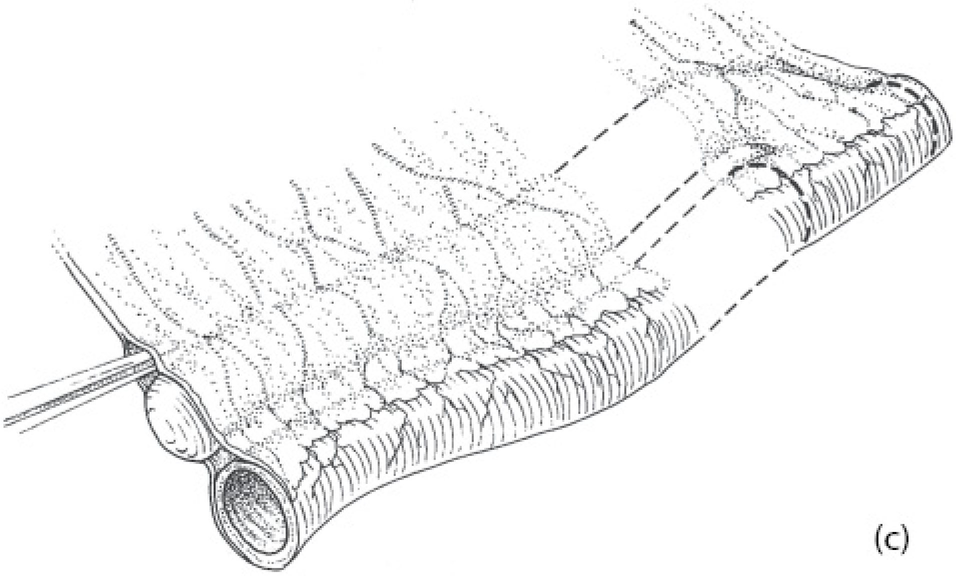
Duplications that are separate from the intestine and located within the mesentery can be removed by careful separation of the two leaves of mesentery, division of the vessels on one side, and enucleation of the duplicated bowel (**Figure 39.9a–c**).



(a)



(b)



(c)

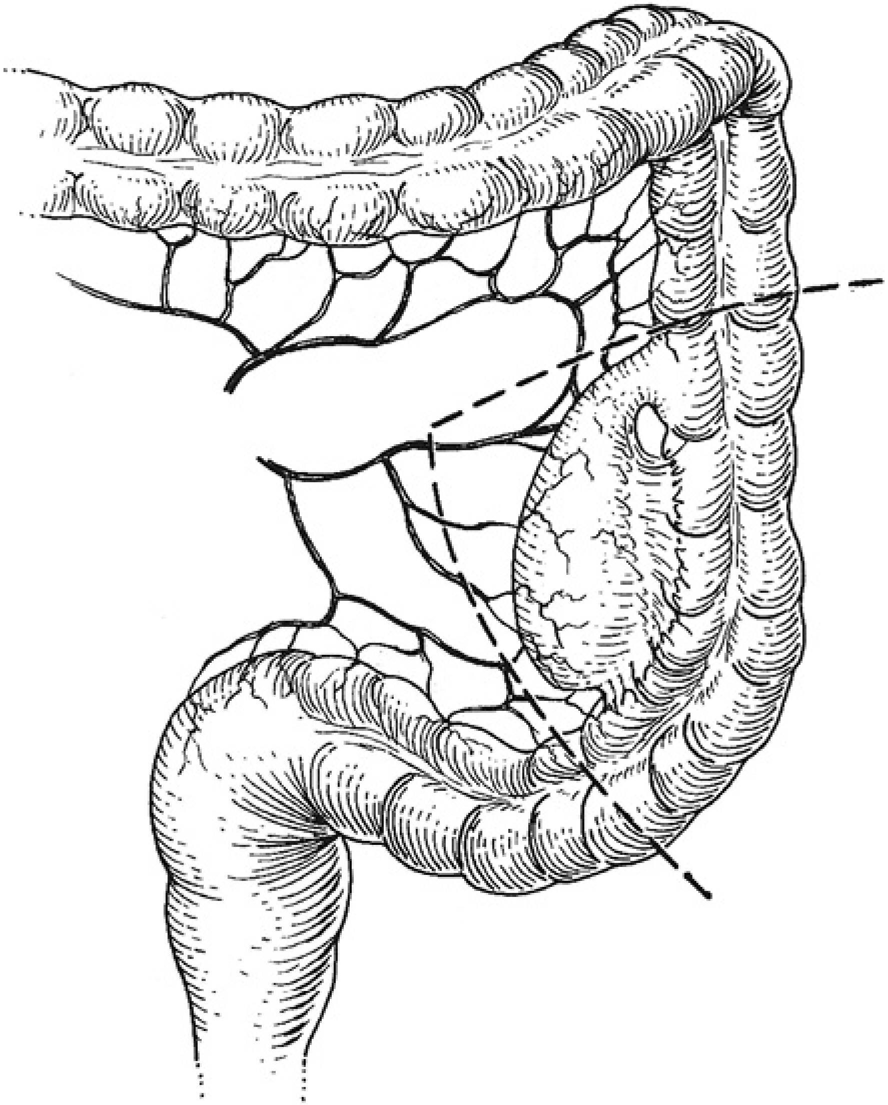
39.9a–c

Short tubular duplications can be excised in continuity with adjacent intestine, but care should be taken to obtain complete excision at the proximal and distal ends of the lesion, where the distinction between normal and duplicated bowel is difficult to detect. Very long tubular duplications, where the remaining intestinal length may be insufficient if completely excised, pose a greater surgical challenge. Treatment possibilities include the following:

- Submucosal resection may be possible, where the mucosal lining is stripped using multiple longitudinal seromuscular incisions.
- If a proximal communication exists, establish a distal communication by fenestration of the common wall to prevent a blind loop. In many cases, this will be the best option, as the shared blood supply makes resection very difficult.
- No surgical treatment and close follow-up may be appropriate if the patient is asymptomatic and no communication exists between the cyst and the bowel.

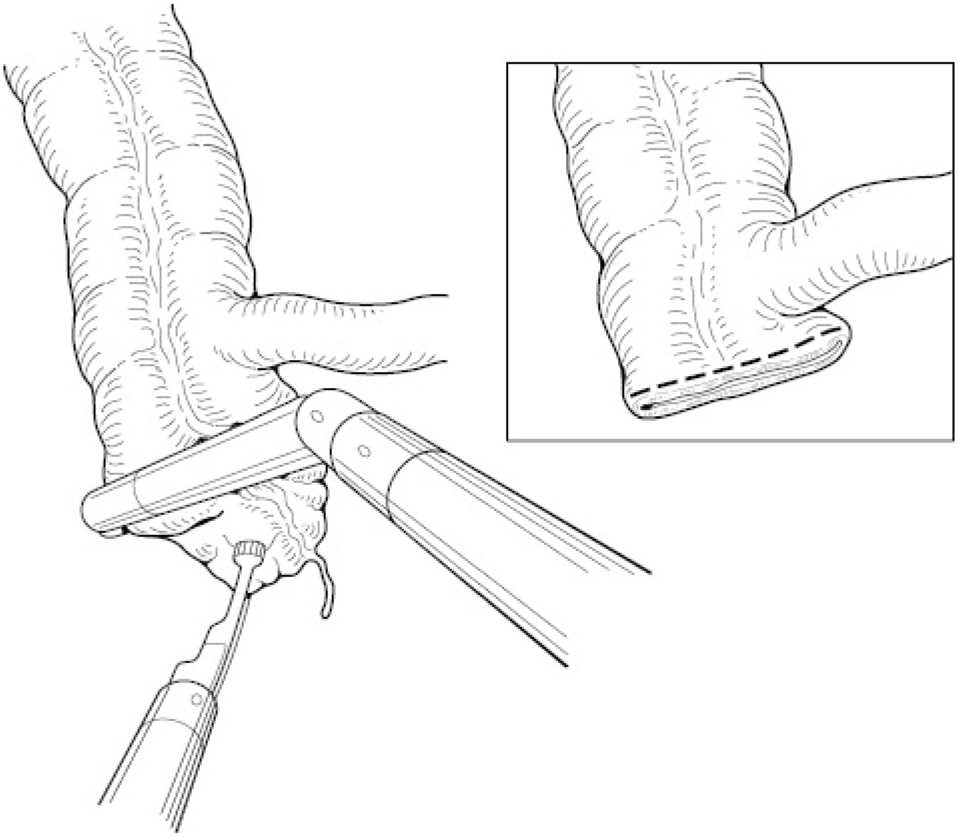
Colonic and appendiceal duplications

Cystic duplications and most short tubular duplications may be excised directly (**Figure 39.10**). The colon is then repaired with an end-to-end anastomosis.



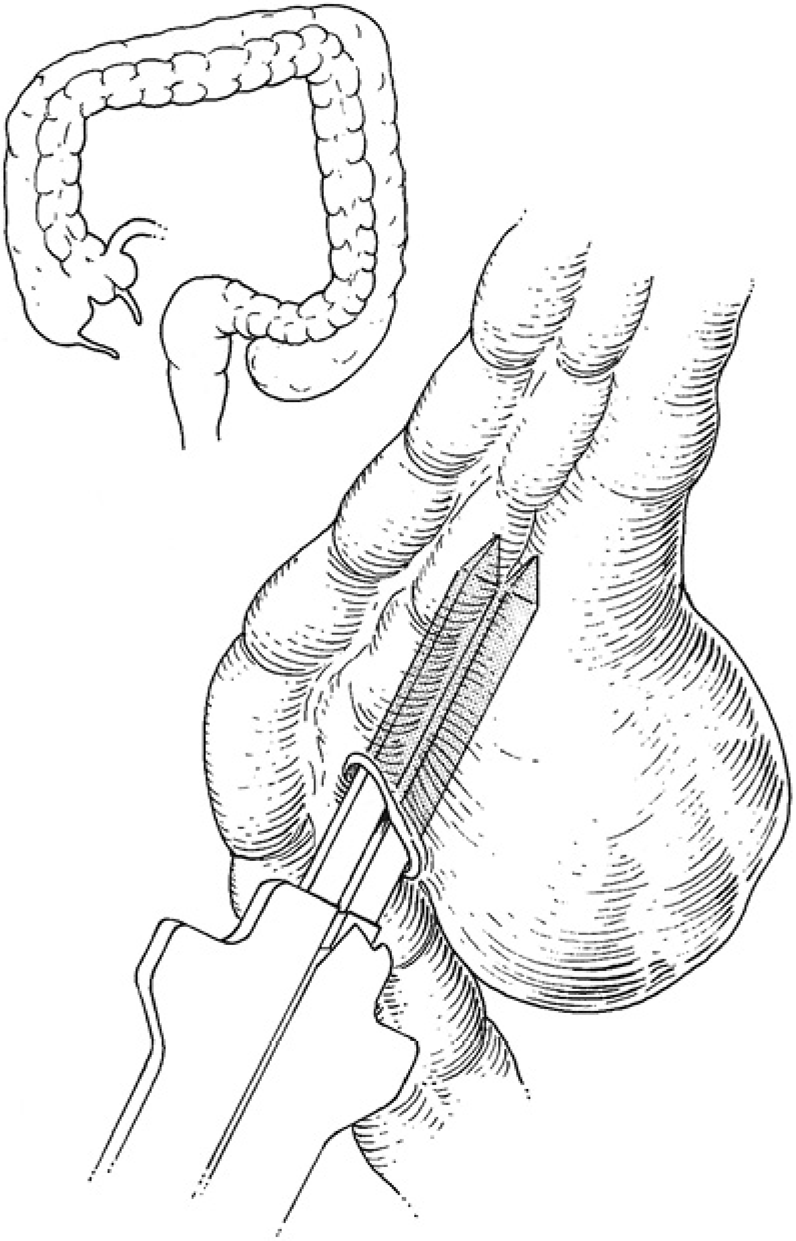
39.10

The operation can also be performed using a laparoscopic or laparoscopic-assisted technique (**Figure 39.11** and see “Minimally invasive surgery for duplications”).



39.11

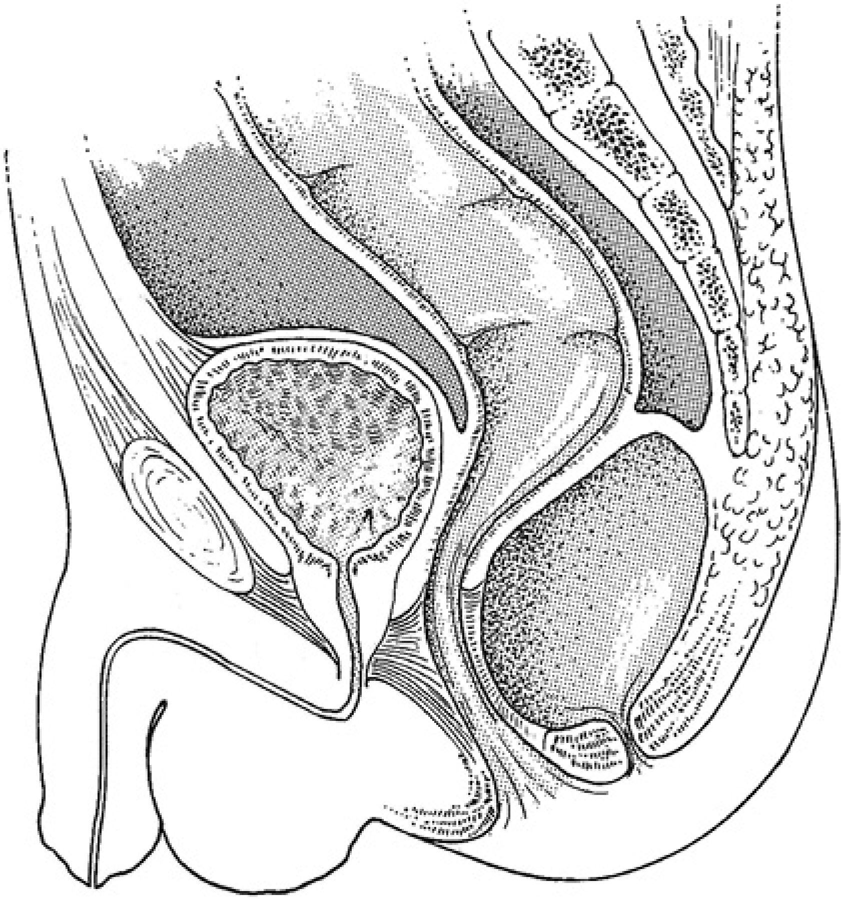
Total colonic duplications are long tubular duplications involving the entire colon. They are located on the anti-mesenteric border or medial to the normal colon and not in the mesentery. The duplication usually communicates proximally with the normal bowel. If the duplication also communicates distally and involves most of the colon and rectum, no treatment is necessary. If the tubular duplication does not communicate distally, a communication must be established, and if a small communication is present, this opening may need to be enlarged. Fenestration can be established by excising a piece of the common wall (**Figure 39.12**).



39.12

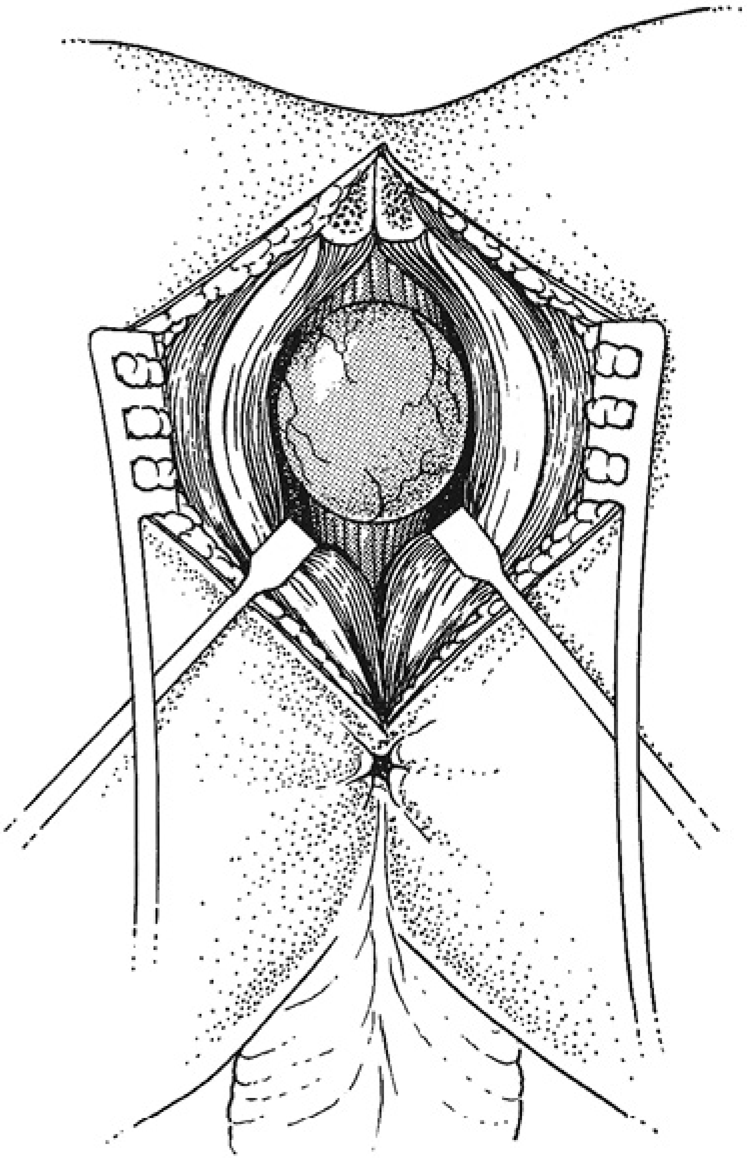
Rectal duplications

Rectal duplications are cystic and located in the retrorectal space. There is often a fistula to the anal canal or perineal region (**Figure 39.13**). The presentation may include constipation, rectal abscess, rectal bleeding, prolapse of the rectum, urinary tract infection, and hemorrhoids. If the cyst is infected, it should initially be drained and then later excised when the inflammation has resolved. Rectal duplication has been associated with a number of other anomalies, including spinal abnormalities, urethral duplication, and anorectal malformations. The abscess can be drained much like any pelvic abscess, i.e. transrectally, transgluteally, or transabdominally, depending on location.



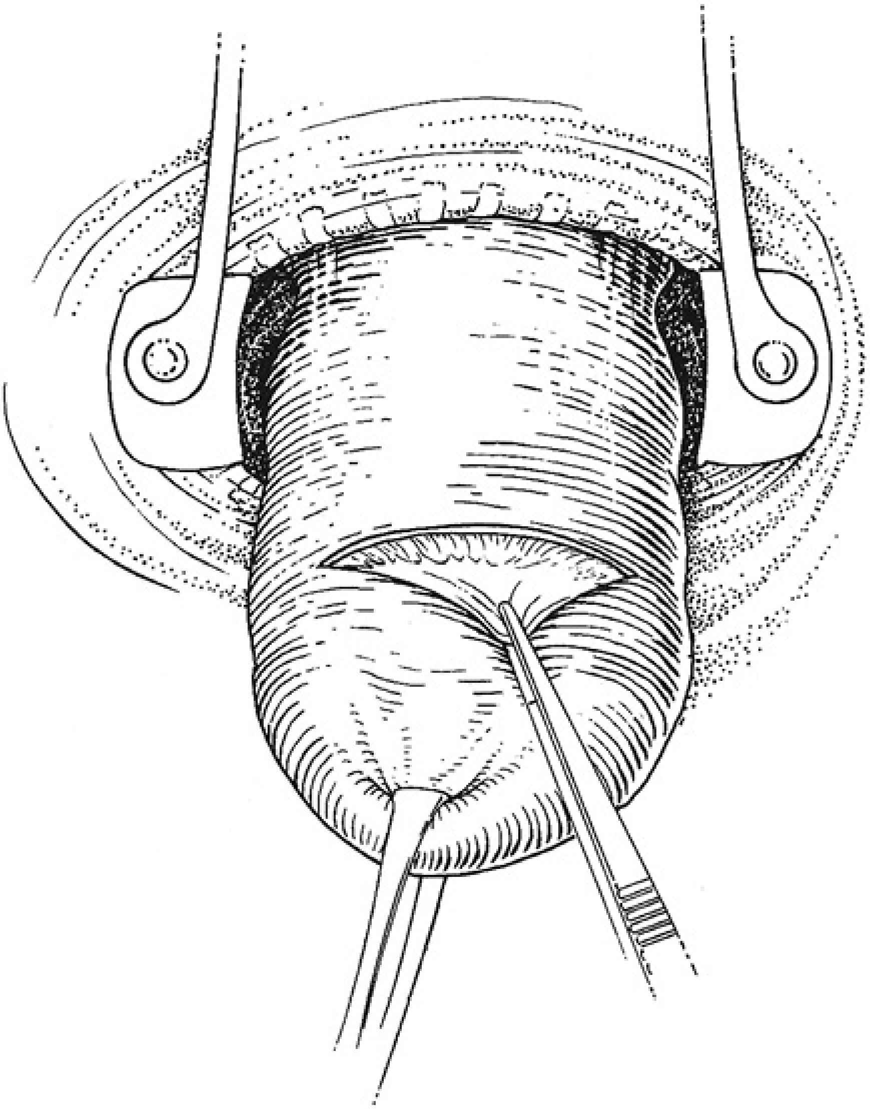
39.13

Treatment options include transanal or posterior sagittal exposure and excision, or transanal fenestration of the common wall if the cyst is large (**Figure 39.14**). Fenestration is a good alternative in cases where the cyst is infected or a fistula between the cyst and the rectum is already present. All patients should receive bowel preparation prior to the operation.



39.14

Small submucosal rectal lesions can be excised through a small incision in the mucosa over the cyst (**Figure 39.15**). To make the dissection easier, the cyst should preferably not be aspirated until the end. After excision, the mucosa is closed with absorbable sutures.



39.15

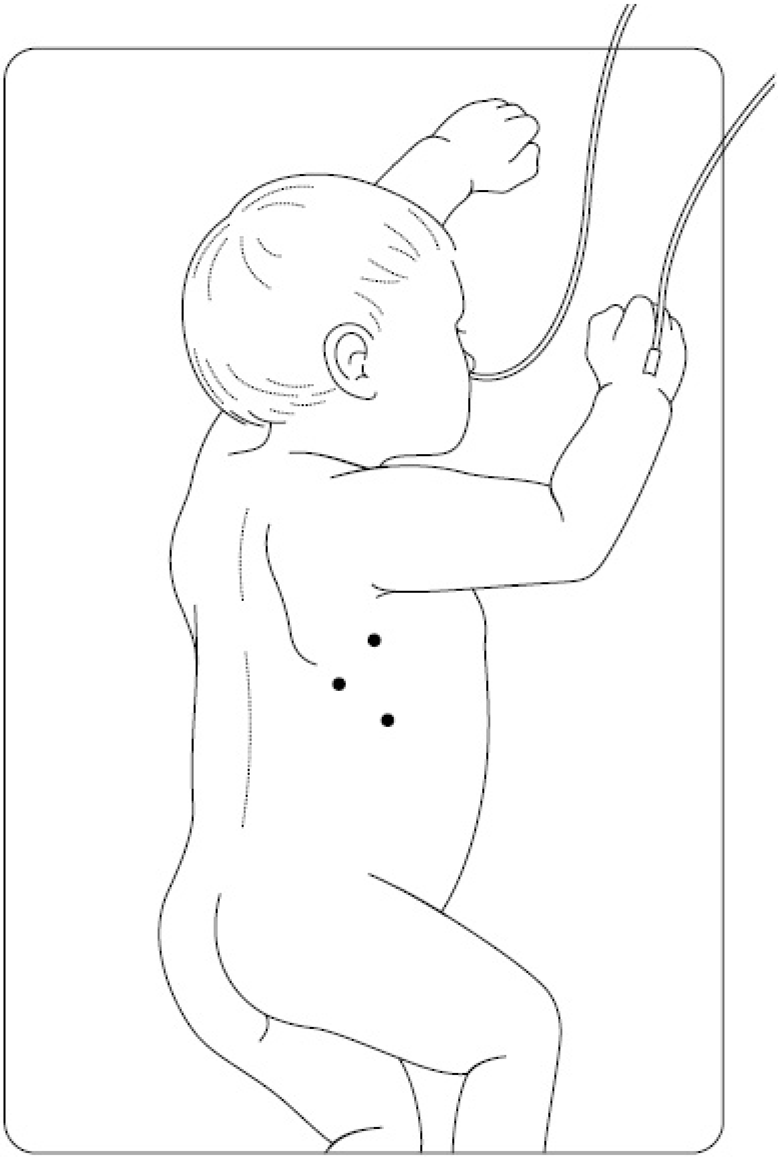
In most cases, rectal duplications can be safely excised without a defunctioning colostomy.

Minimally invasive surgery for duplications

THORACIC

Esophageal duplications are often amenable to thoracoscopic resection. Thoracoscopy minimizes postoperative pain, decreases hospital stay, and provides a better cosmetic result. Most commonly, the esophageal duplications are non-communicating, intramural, cystic lesions located on the right side of the esophagus, although they may also occur on the left side. The surgical approach should use whichever side gives the best exposure to the cyst. When deciding on an open versus thoracoscopic approach, it is important to rule out an extension to the spinal canal, which would necessitate an open approach.

The child is positioned in a semi-prone position at approximately 30 degrees; this gives the best access and exposure of the posterior mediastinum (**Figure 39.16**). The majority of esophageal duplications are approached through the right chest due to their location. It can be beneficial to use a bronchial blocker to minimize ventilation of the right lung. It is best not to drain the duplication cyst during the dissection. It can be drained before removal from the chest to avoid an unnecessary large incision.



39.16

The first port is placed in the mid-axillary line in the fourth or fifth intercostal space. The remaining two ports can be placed under direct visualization, taking into consideration the location of the esophageal duplication. The middle port is placed in the posterior axillary line, while the lower port is placed in the anterior or mid-axillary line.

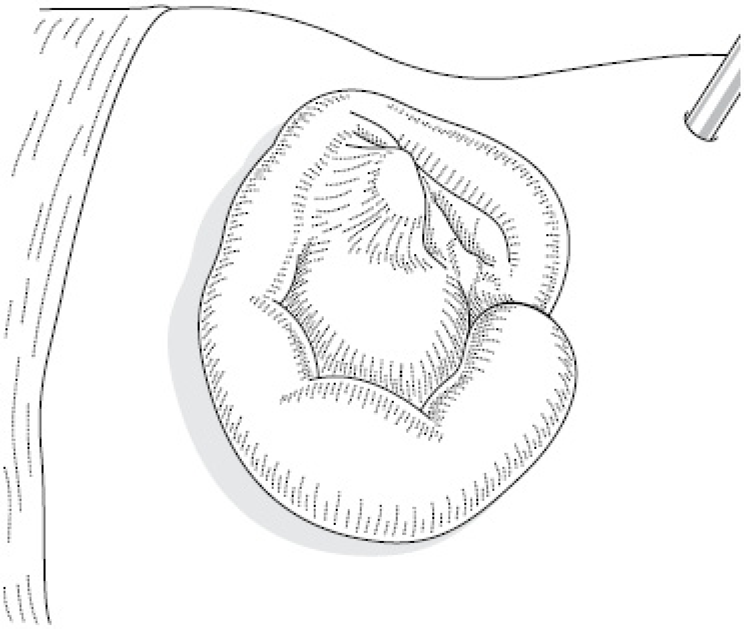
Esophagoscopy may be necessary to locate the lesion. A large orogastric tube or a Maloney dilator should be placed prior to starting the dissection to help with identifying the esophageal wall. Injury to the thoracic duct is a potential pitfall, especially if the duplication is located low in the chest on the right side.

The dissection of the lesion is performed with a combination of sharp dissection with hook cautery and gentle blunt dissection. The principles for the dissection are the same as for an open resection. If there is a weakness of the wall where the duplication was excised or if the esophagus was entered, this can be closed with several laparoscopic sutures. A chest tube is not necessary in most cases, but one can be placed through the lower trocar site at the discretion of the surgeon.

ABDOMINAL

Most duplications in the abdomen can be approached using laparoscopic techniques or a laparoscopic-assisted approach. Initially, an umbilical port is placed. This is followed by the placement of two or three additional working ports, usually 3 mm or 5 mm. A 12 mm port might be necessary depending on the endoscopic linear stapler that is used, and this port can also be used for removal of the specimen. If it is anticipated that a laparoscopic-assisted approach is necessary, one port site is placed over the location of the duplication so that it can be used as part of the open incision.

For resection of a small bowel duplication and primary anastomosis, it is often best to bring the duplication out through the umbilicus and perform an extracorporeal dissection and anastomosis (**Figure 39.17**). This often requires a short extension of the umbilical incision. Cystic small bowel duplications can be aspirated under direct visualization with a large gauge needle either transabdominally or at the umbilical incision prior to externalization. By aspirating the cyst, the decrease in size will make it possible to use a smaller incision, often just confined to the umbilicus.



39.17

Retroperitoneal duplications and spinal involvement

Retroperitoneal duplications can be very large and difficult to remove. It is important to identify adjacent and involved structures to prevent inadvertent injury during the operation. The precise procedure and exposure are dictated by the location and symptoms of the duplication. Intradural and extradural spinal duplications should always be treated in collaboration with a neurosurgeon.

POSTOPERATIVE CARE

The patient should be monitored and followed up according to the extent of the procedure and the findings during the operation. Patients who are managed by incomplete excision or fenestration and have cyst tissue left behind should be followed closely, with repeat ultrasound examinations on a regular basis.

OUTCOME

Generally, patients with the most commonly found cystic duplications of the small and large intestine have an excellent outcome, with several series reporting no mortality or significant morbidity after surgical management. The same applies to short tubular gastric duplications and isolated rectal duplications. Patients who present with thoracoabdominal duplications, complete obstruction of the gastrointestinal tract, bleeding, perforation, and involvement of the mesenteric vessels often have a more difficult clinical course. Associated anomalies, when present, may have a significant impact on long-term outcome.

VIDEO

OPS_8e_video_39.1 Laparoscopic excision of duodenal duplication cyst

FURTHER READING

Azzie G, Beasley S. Diagnosis and treatment of foregut duplications. *Semin Pediatr Surg* 2003; 12: 46–54.

Diamond IR, Teckman J, Langer JC. Laparoscopy in the management of colonic duplication. *Pediatr Endosurg Innovat Tech* 2003; 7: 439–43.

Jeziorczak PM, Warner BW. Enteric duplication. *Clin Colon Rectal Surg* 2018; 31(2): 127–31.

Ladd WE, Gross RE. Surgical treatment of duplications of the alimentary tract: Enterogenous cysts, enteric cysts, or ileum duplex. *Surg Gynecol Obstet* 1940; 70: 295–307.

Norris RW, Brereton RJ, Wright VM, Cudmore RE. A new surgical approach to duplications of the intestine. *J Pediatr Surg* 1986; 21: 167–70.

Stern LE, Warner BW. Gastrointestinal duplications. *Semin Pediatr Surg* 2000; 9: 135–40.

Stringer MD, Spitz L, Abel R, et al. Management of alimentary tract duplication in children. *Br J Surg* 1995; 82: 74–8.

40

Intussusception

MELANIE HIORNS AND JOSEPH I. CURRY

INTRODUCTION

An intussusception is an invagination of one part of the intestine (the *intussusceptum*) into another part of the intestine (the *intussuscipiens*). It most commonly occurs in a proximal to distal direction. The most common area to be affected is the ileum intussuscepting into the cecum and ascending colon. The incidence is highest in infants between the ages of 4 and 10 months, but it can also be found in neonates and adults.

ETIOLOGY

The most common etiological factor in the so-called "idiopathic group" is a preceding viral illness either of the upper respiratory tract or gastroenteritis. The resulting viremia stimulates the gut-associated lymphoid tissue (GALT) with enlargement and edema of Peyer's patches on the luminal surface of the distal small bowel. At the time of laparotomy, these enlarged Peyer's patches can be frequently found at the lead point of an intussusception along with marked lymphadenopathv in the mesentery.

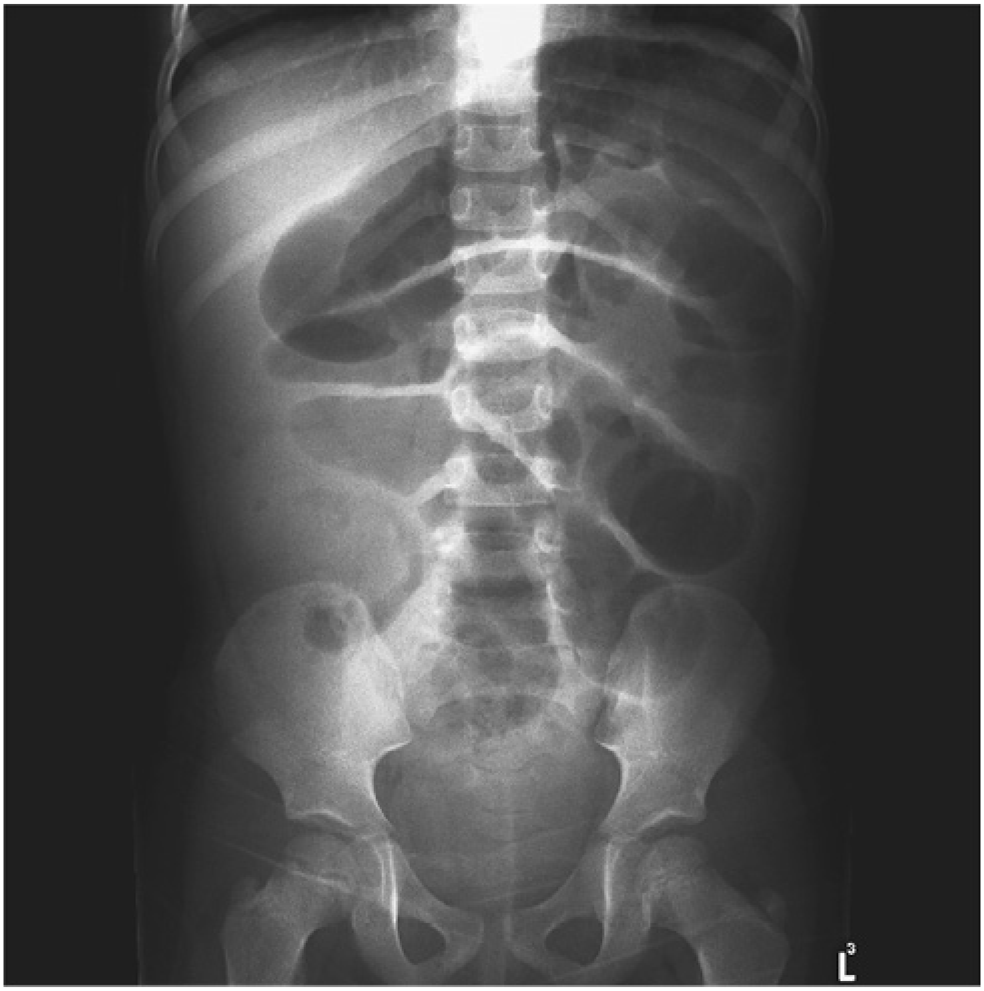
Up to 10% of children will have a pathological lead point, and of these the most common is a Meckel's diverticulum, but polyps, duplication cysts and solid tumors have also been described. A pathological lead point should always be suspected in a child who presents outside the normal range or who has multiple episodes.

Intussusception may also follow administration of rotavirus vaccine (specifically RotaShield□, subsequently withdrawn), though the risk remains unquantifiable. The condition is seen to occur more commonly in boys, unless there is a pathological lead point, in which cases the incidence is equal.

CLINICAL PRESENTATION

Colicky abdominal pain with the infant characteristically "drawing up the legs" is one of the first clinical signs of intussusception. The child will initially be well in between spasms but will later become pale and lethargic. As intestinal obstruction develops, the vomiting will become bilious. Bright red rectal bleeding mixed with mucous, the so-called "red currant jelly stool," will be seen in up to 25% of cases and more frequently if digital rectal examination is performed. A palpable mass is felt in the right upper quadrant or epigastrium along with a distinguishable feeling of emptiness in the right iliac fossa (Dance's sign). Tenderness with evidence of peritonism indicates ischemia or perforation of the intestine. Marked hemodynamic instability with depletion of the intravascular volume can be encountered in children with intussusception. Tachycardia and decreased capillary return should prompt vigorous resuscitation. The diagnosis should always be borne in mind when dealing with a child with shock of unknown cause.

Plain abdominal X-rays are often variable and non-specific and may have an entirely normal appearance. Alternatively, they may show a relative paucity of gas, possibly in the right iliac fossa. As time progresses, the features of small bowel obstruction with dilated gas-filled loops and the soft tissue mass of the intussusceptum may be seen (**Figure 40.1**). The diagnosis should be suspected on clinical grounds and confirmed by ultrasound (US).

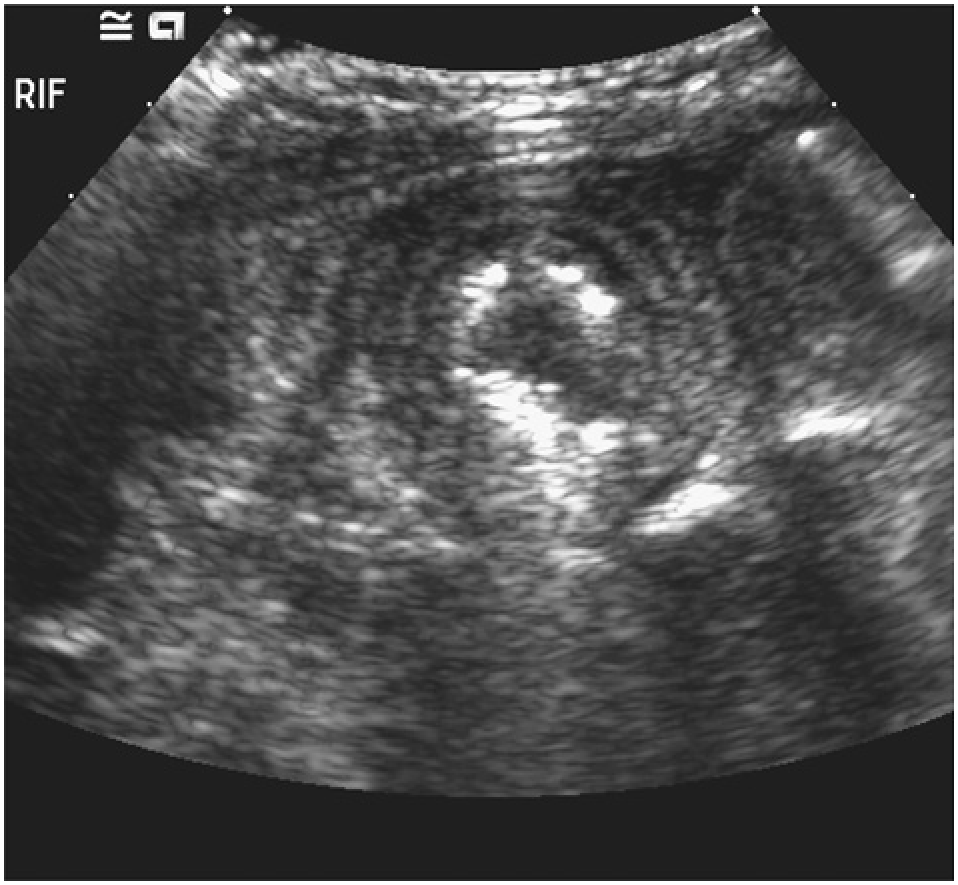


40.1 Plain abdominal X-ray of the abdomen of a child with intussusception showing small bowel obstruction with paucity of gas in the right iliac fossa.

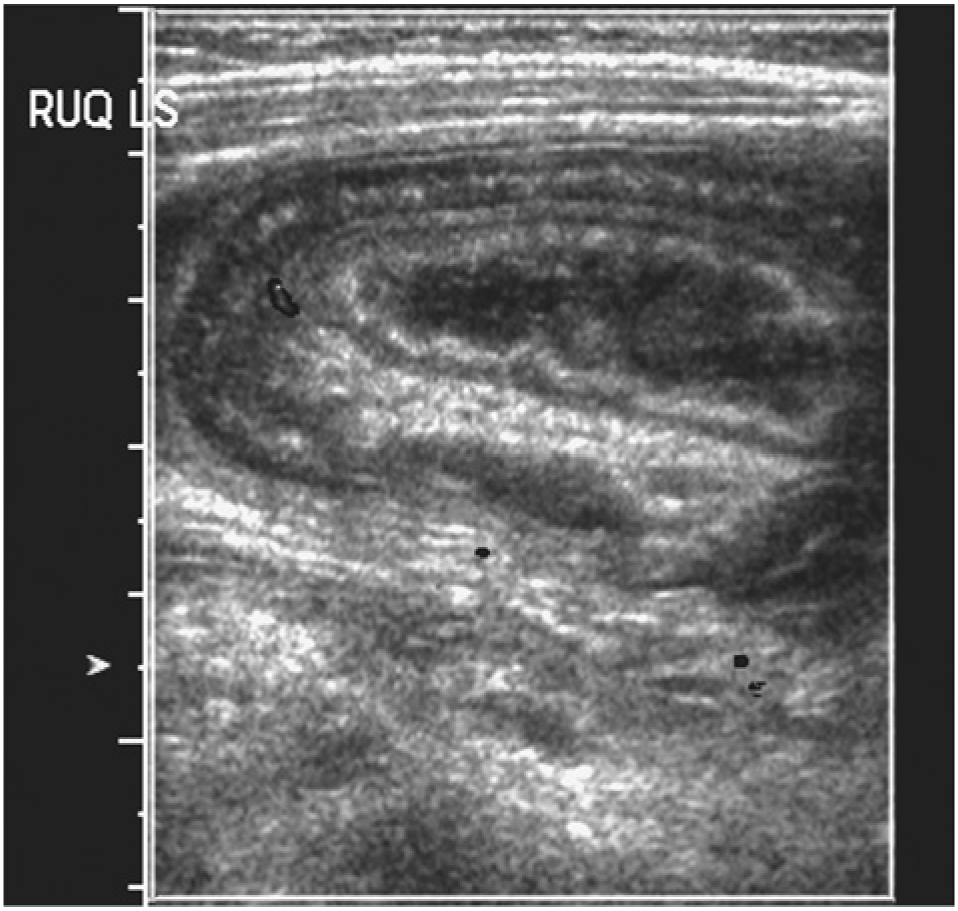
DIAGNOSIS

Ultrasound is highly accurate in the diagnosis with a sensitivity of 98–100% and a specificity of 88–100%. False-negative studies are rare, even for less experienced operators, and therefore the absence of intussusception on US should generally exclude the diagnosis.

The characteristic appearance is a series of concentric rings on the transverse view and of an oval, multi-layered mass on the longitudinal view (**Figures 40.2 and 40.3**), the layers representing the invaginated layers of the bowel wall and edematous mucosa. The most common site is in the right hypochondrium, but it may extend as far down as the rectum. US may not always be able to show the full extent as gas-filled loops of small bowel may impair visualization in the mid-abdomen and the pelvis.

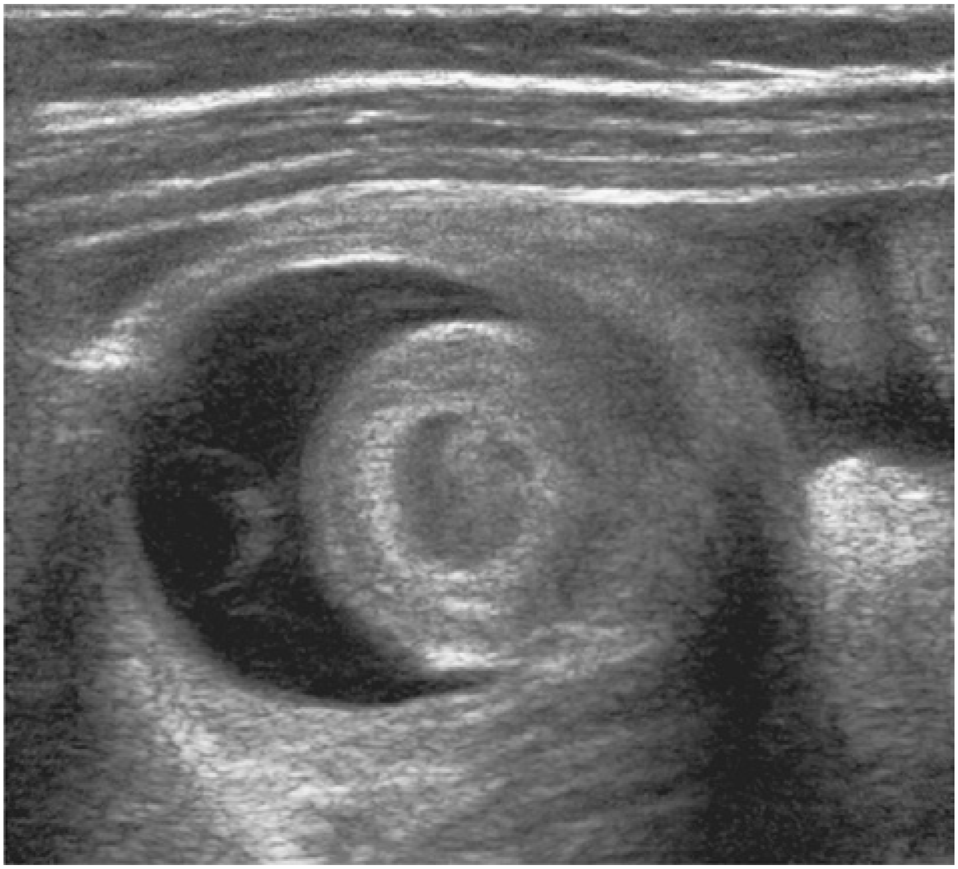


40.2 Ultrasound scan (transverse section) showing the typical "target" sign of intussusception.



40.3 Ultrasound scan (longitudinal section) showing the oval-shaped intussusception ("pseudokidney") with echogenic mucosal walls.

Color Doppler can be used to assess the vascularity of the intussusception. If no color flow is seen, it implies that the intussusception may be of longer duration and may be harder to reduce non-surgically. Absence of color flow does not necessarily imply that it is avascular or necrotic and, all other factors being satisfactory, the child should still have an attempted radiological reduction. Fluid seen between the intussuscipiens and the intussusceptum, so-called "interloop fluid" (**Figure 40.4**), is associated with increased failure of pneumatic reduction. Lymph nodes are a common finding and are usually a reflection of the underlying inflammatory process, but they may represent the lead point, especially if associated with mucosal thickening.



40.4 Fluid seen between the intussuscipiens and the intussusceptum.

US is also useful for assessing the presence and extent of free intraperitoneal fluid. Small amounts of fluid are commonly seen on US, but larger amounts of fluid may indicate bowel perforation. If this is a clinical concern, a plain X-ray may be helpful.

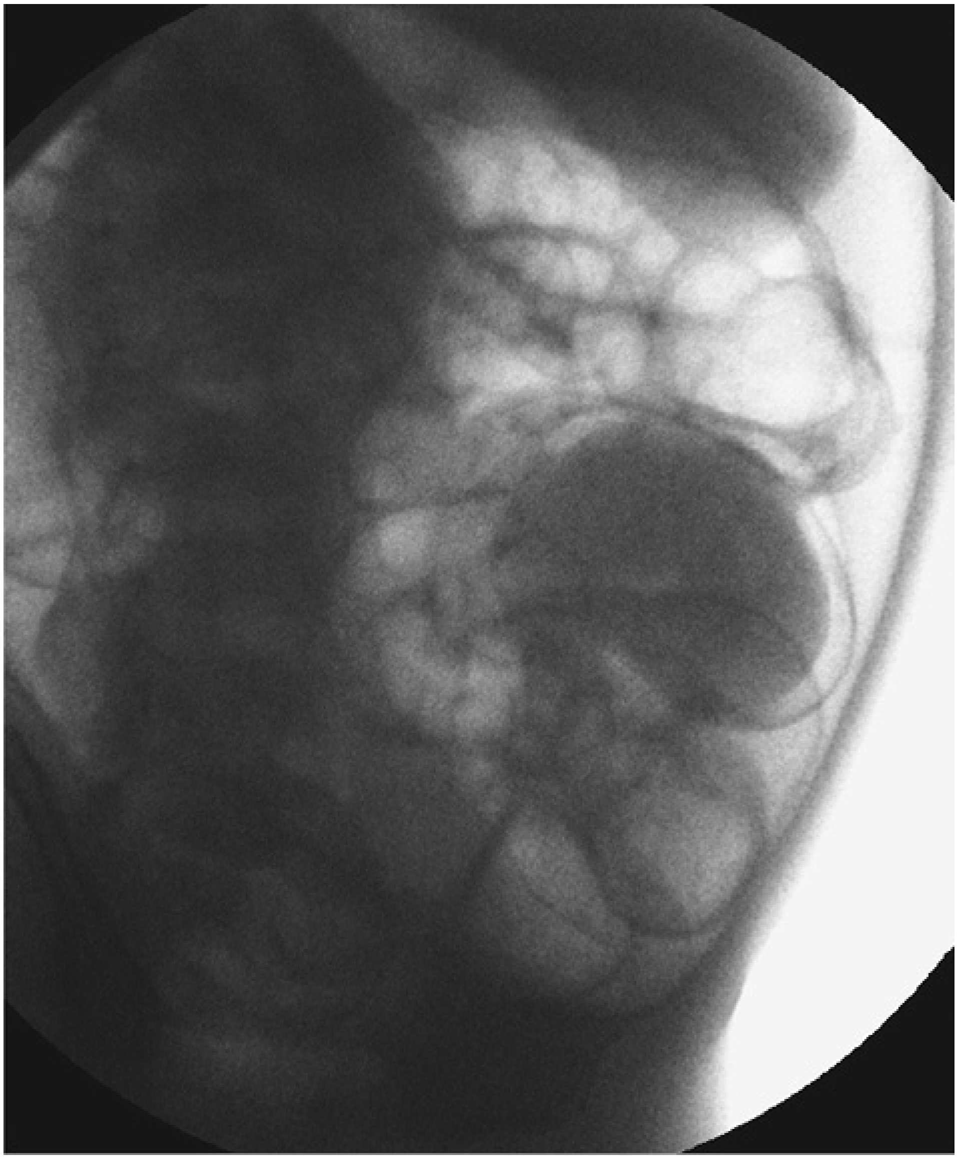
NON-OPERATIVE MANAGEMENT

Once the diagnosis has been confirmed, and after liaison between the surgeon and radiologist, the child would normally proceed to radiological reduction, provided that the child is adequately resuscitated and there are no contraindications, such as free intraperitoneal air or signs of peritonism. A history of more than 24 hours or absent color flow on Doppler are not considered contraindications but may indicate that reduction may be more difficult and should be undertaken with caution. There is no doubt that air enema reduction is more successful with a history of <24 hours. Radiological reduction should be attempted only in centers also offering pediatric surgery in case of complication or unsuccessful reduction.

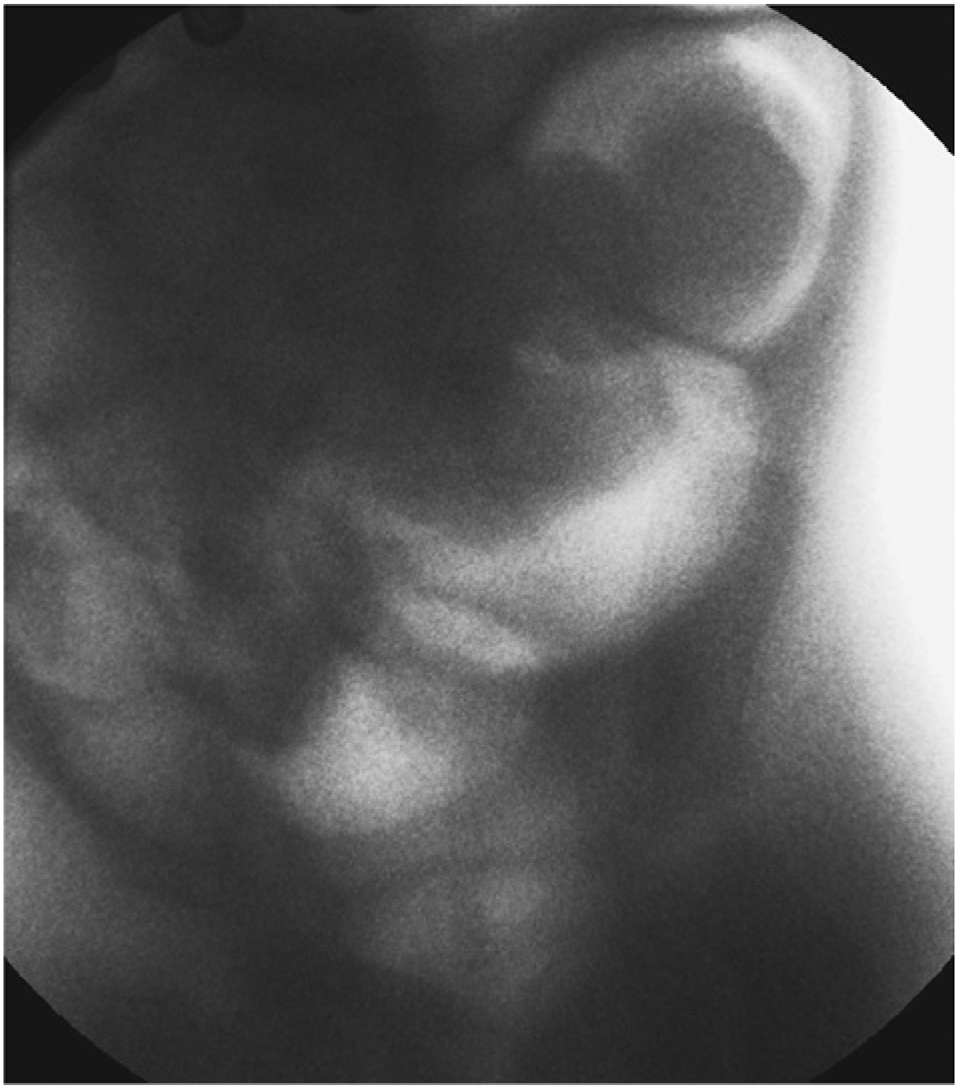
It is important that the child has good IV access and has been fully resuscitated. A nasogastric tube should be passed and prophylactic antibiotics administered. Analgesia can be given at the surgeon's discretion.

Radiological reduction is performed using air under fluoroscopic guidance. It is also possible to use water or air under US guidance. Fluoroscopy allows visualization of the whole abdomen and therefore early detection of perforation, but it uses ionizing radiation. US uses no radiation but can only follow the head of the intussusception. Hydrostatic reduction using barium or water-soluble contrast media is generally no longer used as air reduction has been shown to be safer, to have a higher reduction rate, and to give a lower patient radiation dose.

When using air reduction, a catheter or feeding tube is placed in the patient's rectum and the anus is occluded either by strapping the buttocks tightly together with tape or by the radiologist gripping the buttocks together between the fingers. Some devices are becoming available to occlude the anus, but the value of these is not yet proven. The catheter is connected to a device or system that can deliver air at consistent pressure which can be set by the operator. The system must include a safety device to prevent the pressure unexpectedly exceeding that set by the operator. The child may be supine or prone, but it is easier to observe the child if they are supine. A control image is obtained and then air is slowly introduced into the large bowel until the first set pressure is reached, typically 80 mmHg equivalent. This slow start allows visualization of the head of the intussusceptum and prevents it being missed if reduction is very rapid. Multiple attempts will then be made to reduce the intussusception usually for 3 minutes each for three times at three increasing pressures: 80, 100, and 120 mmHg. This will be monitored very closely under fluoroscopic guidance so that, if perforation does occur, it is immediately detected (**Figure 40.5**) and the procedure can be stopped immediately before a pneumoperitoneum causes splinting of the diaphragm and respiratory arrest, or increased vasovagal stimulation causes a cardiac arrest. The procedure is also terminated if air is seen to track along the sides of the intussusceptum, the "air dissection" sign, as this implies attempted reduction will be unsuccessful (**Figure 40.6**).

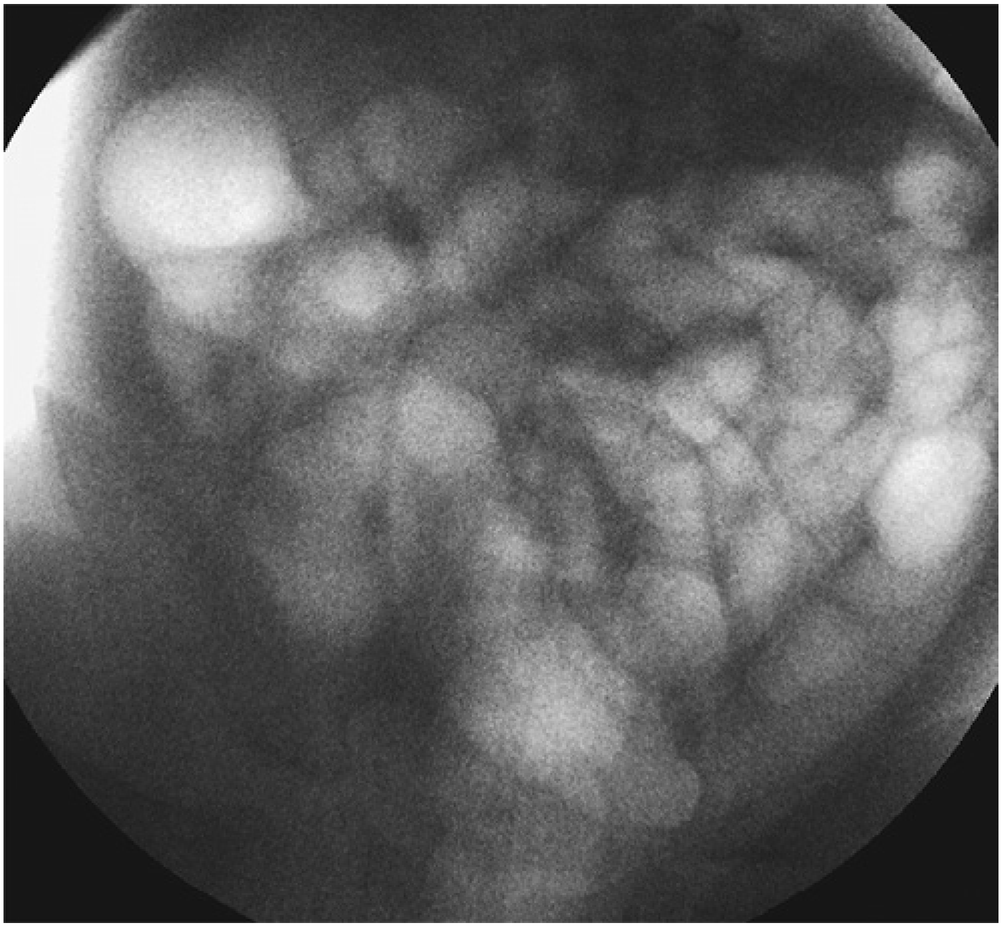


40.5 Pneumoperitoneum occurring during attempted reduction. Free air is seen over the right side of the image.



40.6 Air dissection along the side of the intussusception.

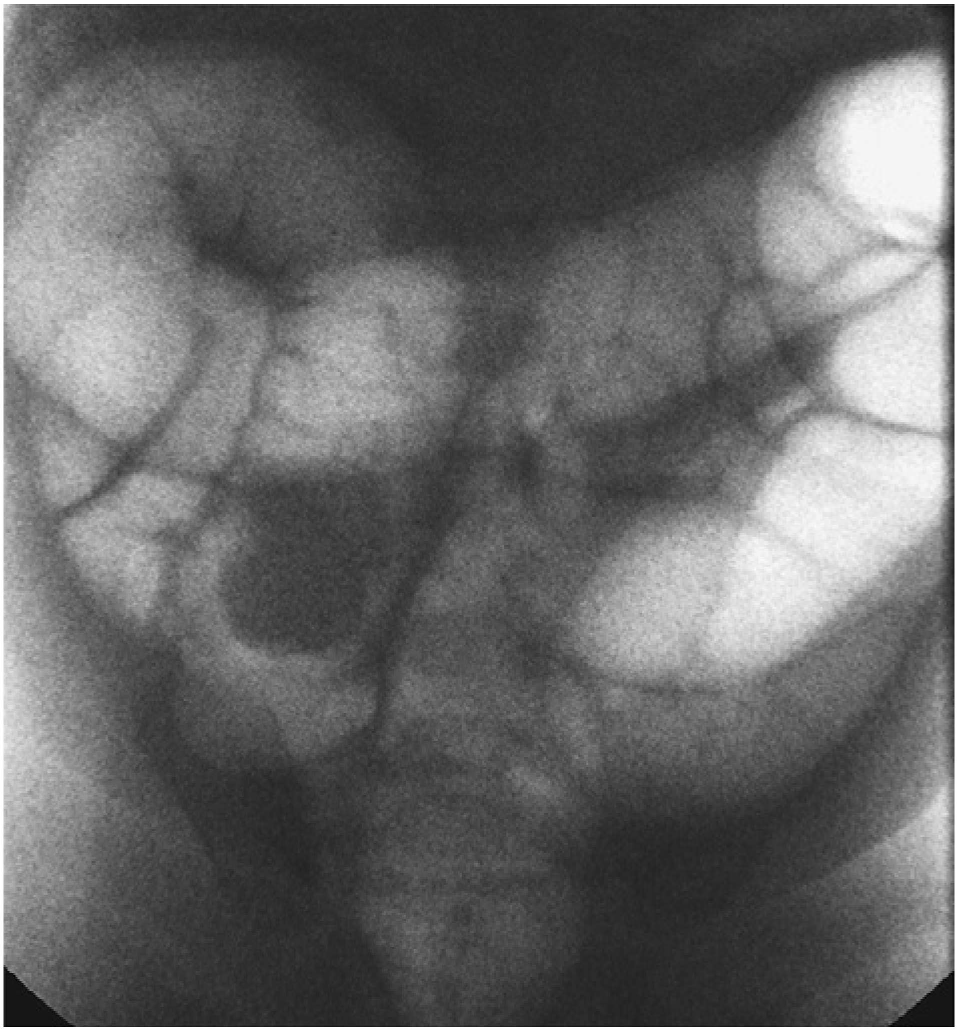
Reduction is achieved when air flows freely into the small bowel (**Figure 40.7**).



40.7 Filling of small bowel with air, indicating successful reduction.

The procedure is often distressing for the child and their parents if they are present in the room. However, only a few centers in the UK give sedation as there is some evidence that, when the child cries and performs a Valsalva maneuver, the brief rise in intra-abdominal pressure assists the reduction while the Valsalva maneuver may provide a protective effect against perforation because external abdominal pressure decreases the transmural gradient. More importantly, if there is a sudden deterioration in the child's clinical condition, this is more likely to be detected in an unsedated child.

If the intussusception is reduced as far as the ileocecal valve but not through it (**Figure 40.8**), it is now considered worthwhile to allow the child to rest for a few hours as edema at the ileocecal valve may settle and a further radiological attempt at reduction can be made to avoid the need for surgery. A second attempt may be made after 4–6 hours if the child is stable and if the intussusception has been partially reduced on the first attempt. Reported rates of 50–80% reduction suggest this may be an important technique in previously failed air enema reductions.



40.8 Persistence of intussusception at the ileocecal valve.

Success rates vary between institutions, but current guidelines suggest that a success rate of at least 70% should be achievable. Perforation rates are typically <2% and the parents should be warned that this would necessitate immediate surgical intervention. A significant pneumoperitoneum requires immediate decompression by the radiologist, usually by the placing of an 18 gauge needle into the abdomen. Perforation with air reduction does not usually result in widespread contamination as the large bowel is usually empty of stool and also because air escapes and rises to the top of the abdomen rather than washing contaminated contents around within the abdomen as fluids would do. Perforation is most likely secondary to bowel necrosis and not excessive pressures.

OPERATIVE REDUCTION

Indications

Indications for operative reduction include:

- Initial evidence of peritonism or perforation
- Perforation during radiological reduction
- Failure of radiological reduction
- Third-time presentation (presentation well beyond the usual age range)

Preoperative preparation

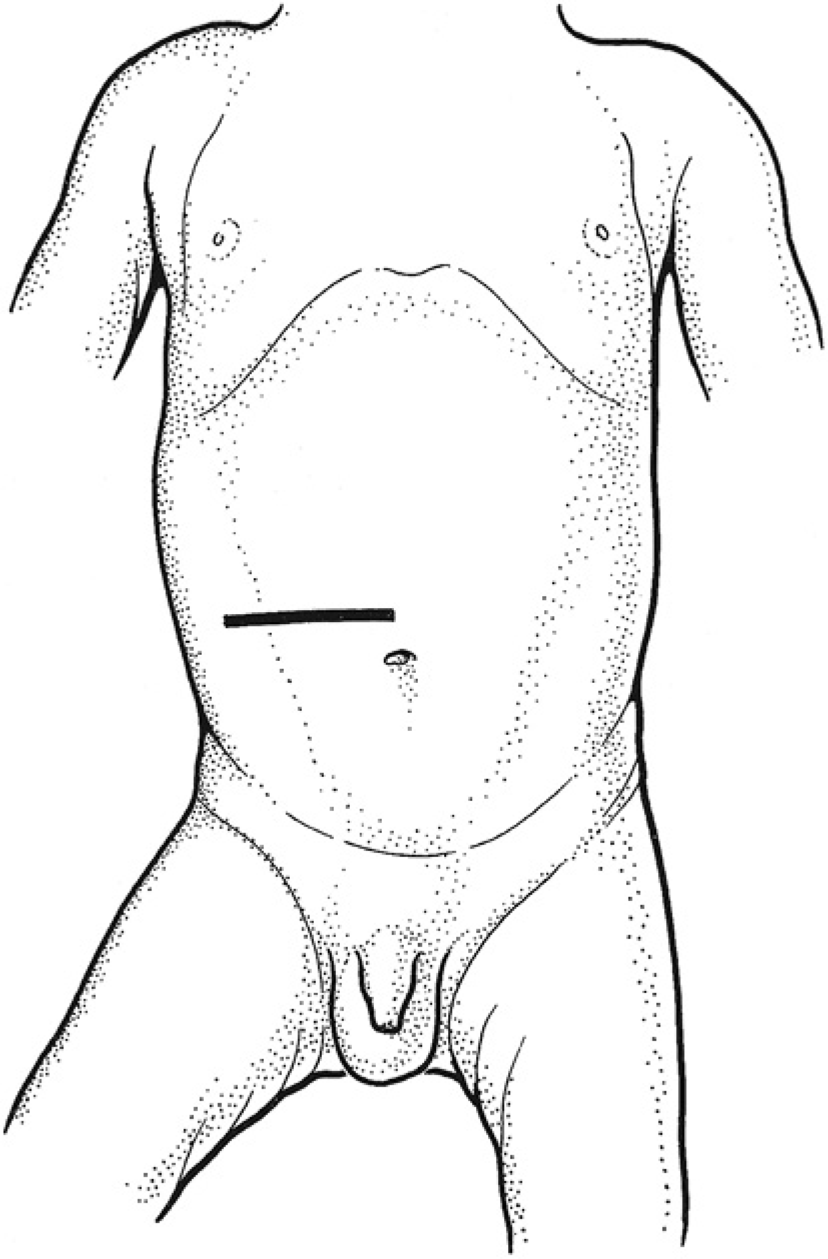
Arrangements should be in place for emergency surgery in all infants undergoing attempted radiological reduction to minimize delay.

Fluid resuscitation should continue and the infant's hemodynamic status be regularly assessed. Broad-spectrum antibiotics should be administered if not previously given prior to attempts at radiological reduction, and a nasogastric tube is passed.

Open operation

INCISION

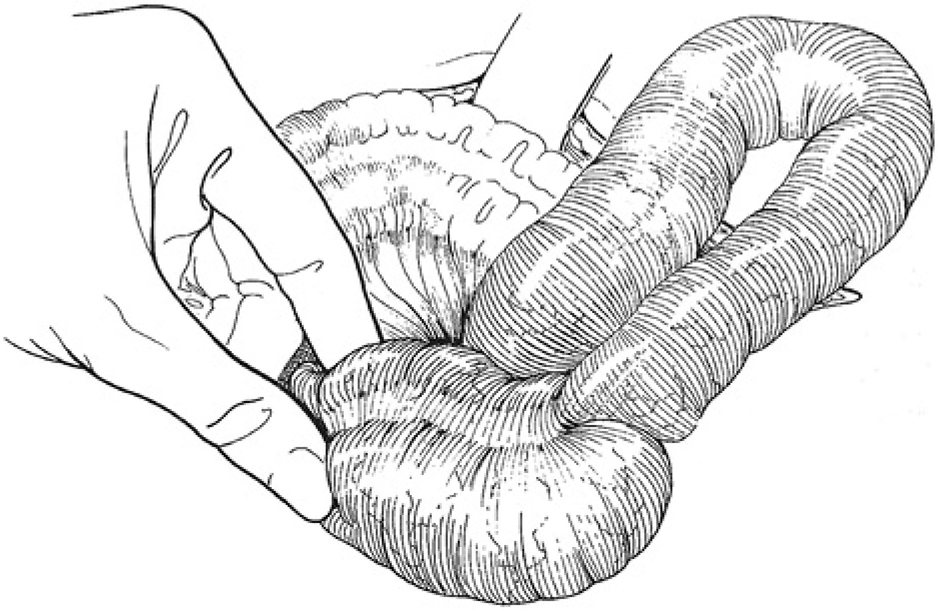
A right transverse skin incision is made either above or below the level of the umbilicus, depending on the presence of a mass or radiological indication of the site of the intussusception (**Figure 40.9**). The lateral abdominal muscles, rectus sheath, and rectus muscle are divided. A sample of the peritoneal fluid is sent for microbiological analysis.



40.9

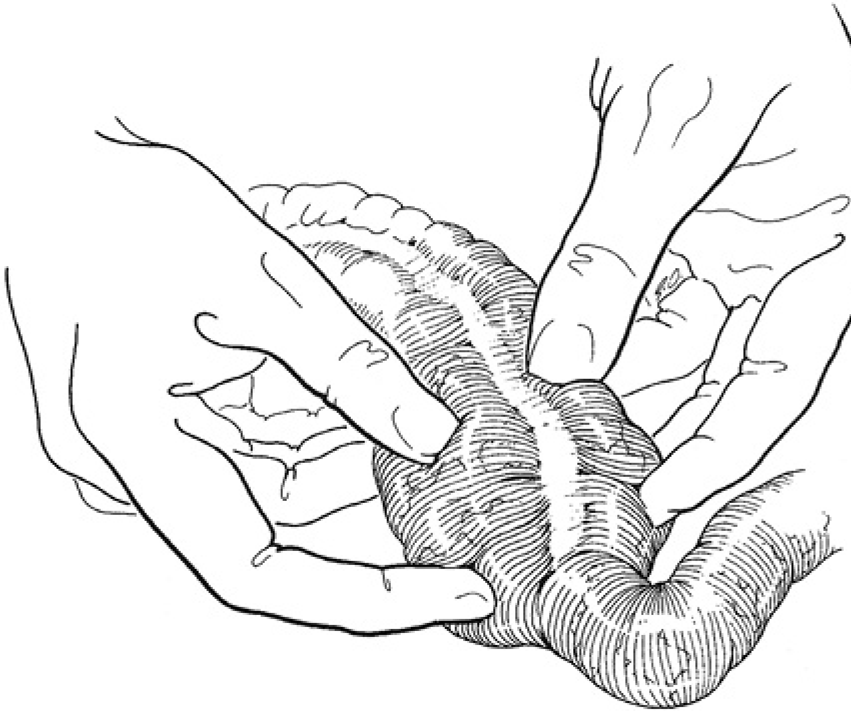
REDUCTION OF INTUSSUSCEPTION

The affected bowel is delivered from the abdominal cavity to facilitate reduction (**Figure 40.10**). This often involves division of the peritoneal attachment of the right colon and cecum using sharp dissection. Once the affected segment has been delivered, all other parts of the intestine are returned to the abdomen. Reduction is achieved by gently applying pressure on the apex of the distal part of the intussusception. Grip on the intestine can be facilitated by the use of a gauze swab. Traction on the proximal intestine should be avoided but a gentle pull may establish the direction in which to apply the reducing squeeze.



40.10

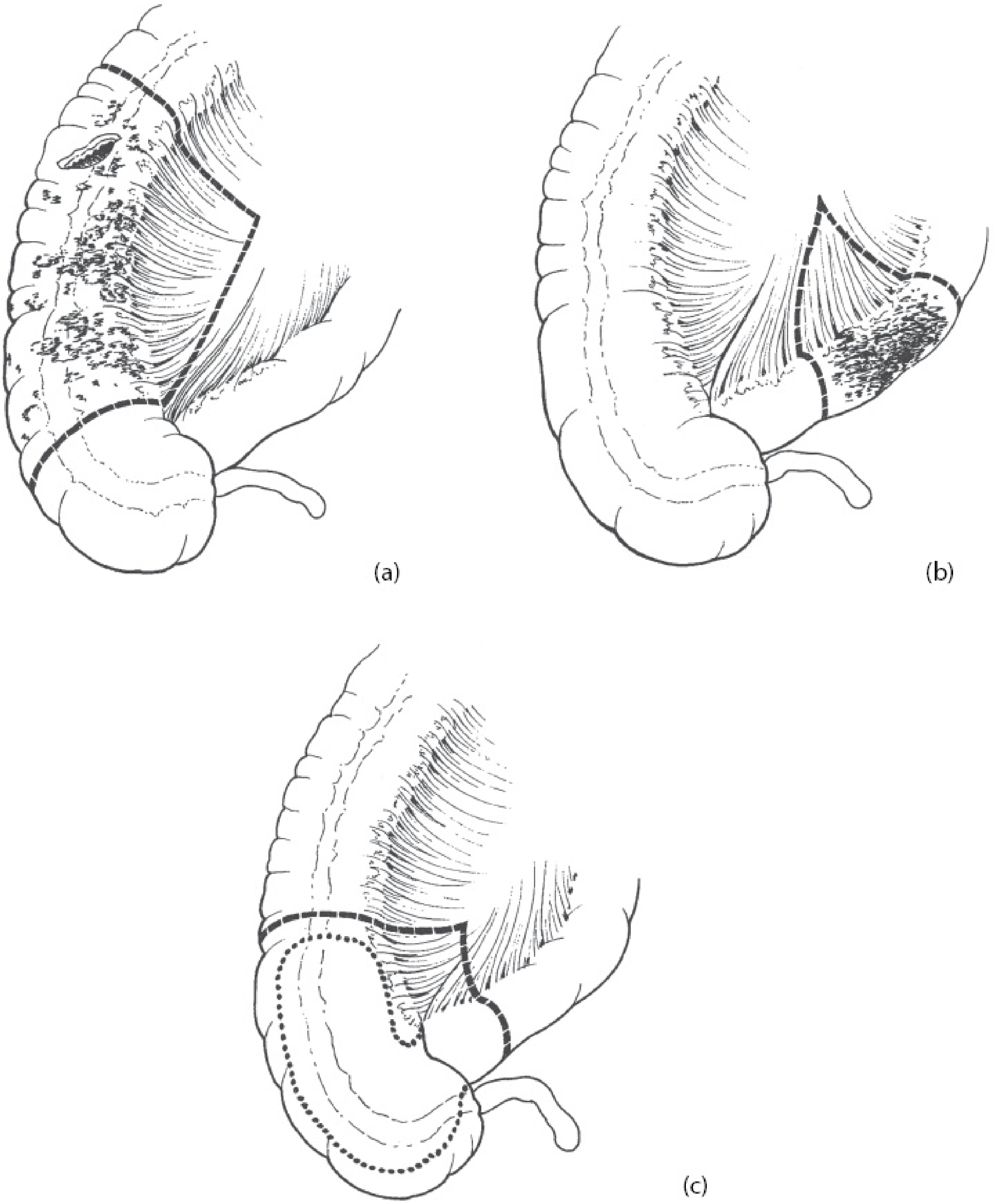
Reduction of the ileum through the ileocecal valve requires patience. Forefingers and thumbs are used to squeeze the apex of the intussusception gently while pulling back the cecal wall (**Figure 40.11**). The gut should be palpated to rule out a pathological lead point, bearing in mind that an edematous ileocecal valve or Peyer's patch can mimic an intraluminal mass.



40.11

RESECTION

Resection will be necessary if the intussusception cannot be reduced, if there is necrotic or compromised bowel after reduction, or if there is a pathological lead point. The intussusception should be reduced as far as possible to minimize the extent of any resection (**Figure 40.12a and b**). This is usually segmental and rarely extends to formal right hemicolectomy.



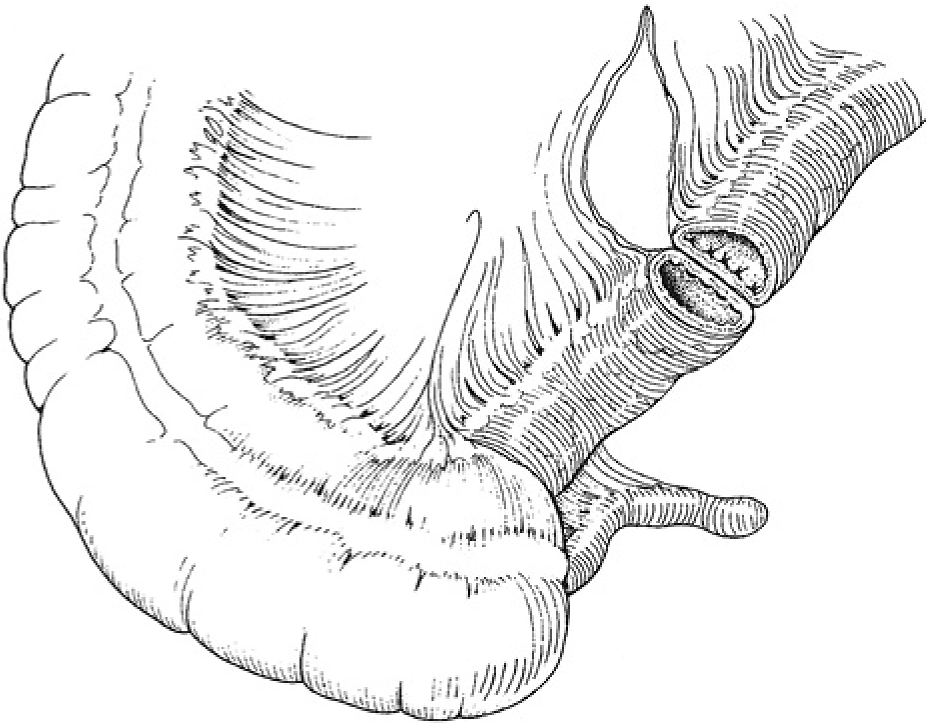
40.12a–c

If the intussusception is extensive, i.e. beyond the splenic flexure, then consideration should be given to examining the duodenojejunal flexure for coexistent malrotation. If present, a formal Ladd's procedure should also be performed.

In the presence of a transverse incision below the level of the umbilicus, it is acceptable to perform an incidental appendicectomy provided the adjacent cecal wall is sound (**Figure 40.12c**).

ANASTOMOSIS

Suction of the proximal and distal bowel avoids the need for bowel clamps. Primary anastomosis is almost always possible (**Figure 40.13**) and is performed with a single layer of interrupted extramucosal sutures with knots placed on the serosal surface (e.g. polydioxanone suture [PDS□]). Any mesenteric defect is closed in a similar fashion. The need for expeditious surgery in a very sick infant, along with doubt about the viability of the resection margins, may require the formation of adjacent stomas. Closure is performed when the infant's condition has improved and the stomas are healthy.



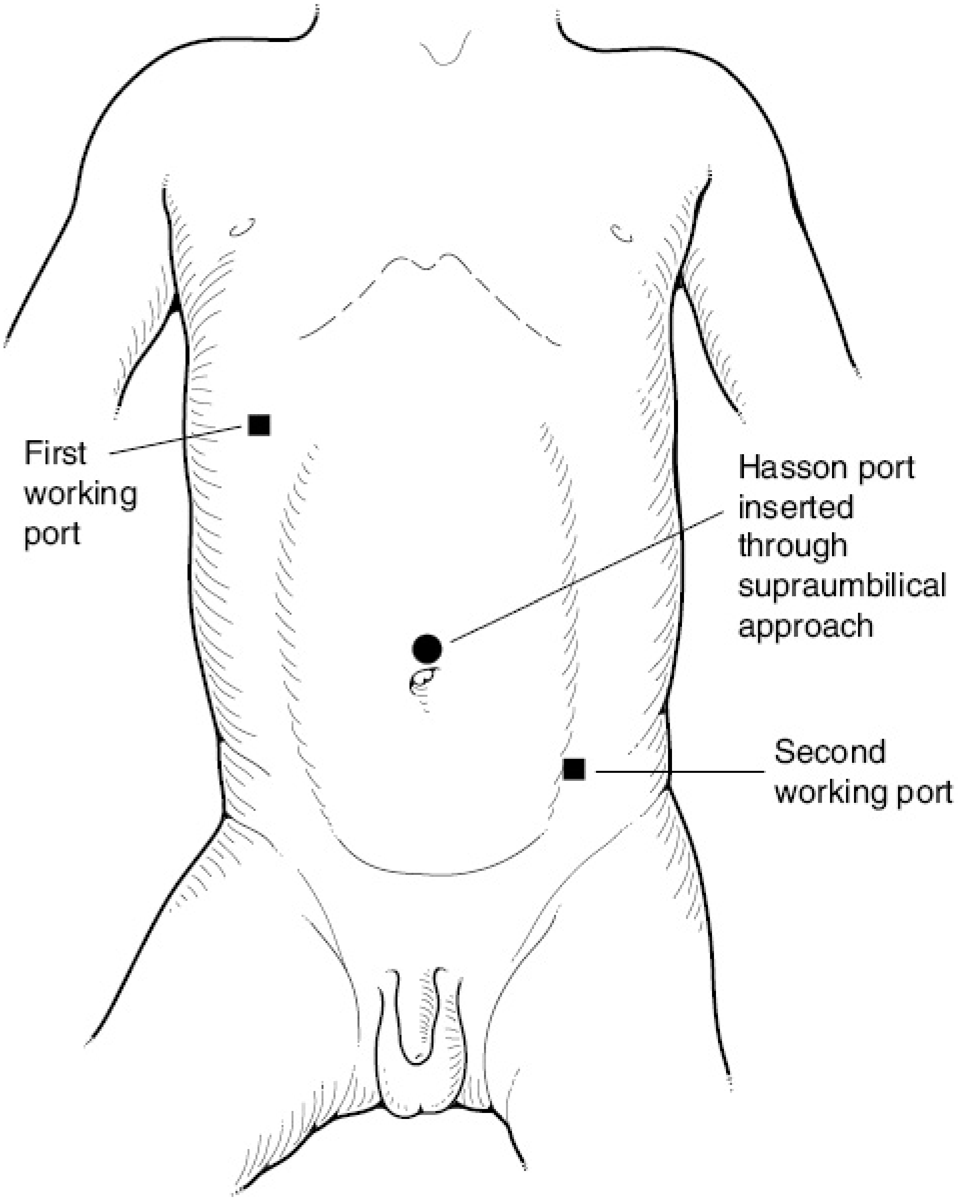
40.13

It may be possible to oversew a perforation using interrupted extramucosal sutures provided the edges are clean and viable.

Laparoscopic reduction

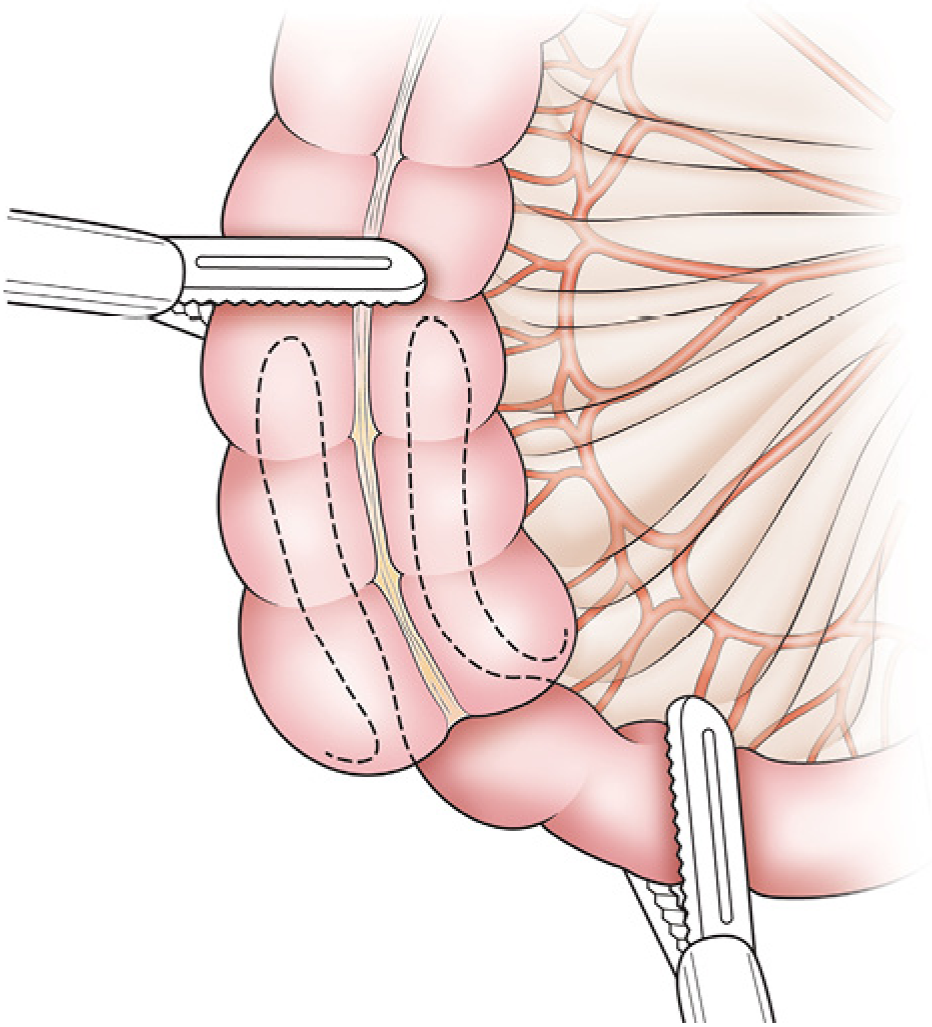
Intussusception is a reasonable indication for minimally invasive surgery. Potential advantages include the ability to diagnose full reduction when this is unclear after pneumatic reduction thus avoiding full laparotomy or cases of recurrence. Reduction itself is often more problematic, due usually to the resulting small bowel dilatation, but if achievable, avoids the trauma of open access and significantly reduces the risk of intraperitoneal adhesions. Irreducible intussusceptions can be externalized through extension of the umbilical port or through a small and accurately placed incision.

Pneumoperitoneum is raised according to surgical preference. Access ports are inserted in the right upper quadrant and left lower quadrant, at right angles to the small bowel mesentery (**Figure 40.14**).


First working port
Hasson port inserted through supraumbilical approach
Second working port

40.14

Attention is initially focused in the right iliac fossa. The small bowel and colon are examined to find the area of intussusception. Reduction is effected by a combination of taxis and traction using atraumatic graspers (**Figure 40.15**). Careful visualization of the bowel is necessary to avoid rupture and intraperitoneal contamination. In the case of successful reduction, the appendix should be left *in situ* as there is no scar in the right iliac fossa.



40.15

The lack of tactile clues to a pathological lead point should be remembered and consideration given to delivering the small bowel through the umbilical port after successful reduction in older children.

POSTOPERATIVE CARE

Appropriate IV fluids and nasogastric decompression are continued until diminution of aspirated content suggests return of intestinal function. Oral fluids are commenced within 24 hours, but in the presence of resection may be delayed a further 24 hours. Continued antibiotic therapy is dictated by surgeon's preference and the presence of peritoneal contamination at the time of laparotomy.

A high temperature is common in the first 24–48 hours postoperatively and usually subsides without specific treatment.

OUTCOME

Recurrences following pneumatic reduction are reported in up to 10% of cases. Further radiological reduction should be attempted, but a third should raise the possibility of a pathological lead point.

Deaths from intussusception have been reported and relate to failure to make the diagnosis and inadequate resuscitation.

Rarely, excision of the ileocecal valve may predispose to anemia secondary to depletion of vitamin B12 or the development of gall stones from loss of bile salts.

FURTHER READING

Burnett E, Parashar U, Tate J. Rotavirus vaccines: Effectiveness, safety, and future directions. *Paediatr Drugs* 2018; 20: 223–33.

Chew R, Ditchfield M, Paul E, Goergen SK. Comparison of safety and efficacy of image-guided enema reduction techniques for paediatric intussusception: A review of the literature. *J Med Imag Radiat Oncol* 2017; 61: 711–17.

Edwards EA, Pigg N, Courtier J, et al. Intussusception: Past, present and future. *Pediatr Radiol* 2017; 47: 1101–8.

Fiegel H, Gfroerer S, Rolle U. Systematic review shows that pathological lead points are important and frequent in intussusception and are not limited to infants. *Acta Paediatr* 2016; 105: 1275–9.

Gartner RD, Levin TL, Borenstein SH, et al. Interloop fluid in intussusception: What is its significance? *Pediatr Radiol* 2011; 41: 727–31.

Kaiser AD, Applegate KE, Ladd AP. Current success in the treatment of intussusception in children. *Surgery* 2007; 142: 469–75.

Sklar CM, Chan E, Nasr A. Laparoscopic versus open reduction of intussusception in children: A retrospective review and meta-analysis. *J Laparoendosc Adv Surg Tech A* 2014; 24: 518–22.

41

Appendectomy

MIKKO P. PAKARINEN

HISTORY

Before the end of the 19th century "appendicitis" was rarely seen, cases typically being described as *typhlitis* (inflammation of the cecum). Inflammation of the appendix was conclusively demonstrated to cause this condition by Reginald Fitz in 1886. The first published report on appendectomy is attributed to Claudius Amyand from England who, in 1735, removed a perforated appendix from the scrotal hernia of an 11-year-old boy. The first deliberate appendectomy (or even appendicectomy) for perforated appendix in a 17-year-old girl was performed by Lawson Tait in Birmingham in 1880 (though published later).

Following this and throughout the 20th century there was a huge increase in prevalence, and it still remains the commonest single reason for an acute surgical abdomen in adults and children. Nonetheless, it continues to be associated with significant morbidity and even occasional mortality. The overall lifetime risk of appendicitis has been estimated as 7–9%.

PRINCIPLES AND JUSTIFICATION

Appendicitis is one of the most common surgical emergencies in children. Prompt appendectomy is the recommended first-line treatment of acute appendicitis, though there have been recent randomized controlled trials in both adults and children showing the possibility of a non-operative approach using only antibiotics.

The indications for surgery are the same for the open and laparoscopic procedures. The latter is associated with a shorter hospital stay in children with uncomplicated appendicitis. Although operation without delay has been the gold standard, there is a trend away from immediate surgery, especially during the night. No increased incidence of complications or perforations has been reported in patients who have been operated <36 hours after admission without clinical signs of complicated appendicitis. Early surgery is still advocated for the youngest children because of increased risk of perforation before surgery.

Appendectomy is also the generally accepted treatment of perforated appendicitis. However, there is significant controversy regarding the treatment of a palpable appendix mass. Some surgeons advocate immediate operation, while others favor initial conservative therapy with IV antibiotics with or without drainage of abscess and interval appendectomy 2–3 months later. Appendicitis with abscess has been considered as a contraindication to laparoscopic surgery. Recent data, however, do not support this; early laparoscopic surgery for an appendix mass is at least as safe as open surgery. Wound infection, hospital stay and bowel obstruction are reduced after laparoscopic removal of a perforated or gangrenous appendicitis whereas the incidence of intra-abdominal abscess increases slightly. Interval appendectomy is typically performed laparoscopically. The need for interval surgery in children has been recently challenged in a randomized controlled trial, suggesting that only a minority of patients develop complications in the following year if left alone.

Appendectomy is also recommended for small (<2 cm) carcinoid tumors at the tip of the appendix, other benign conditions of the appendix, and as part of Ladd’s procedure for midgut malrotation. Incidental removal of a normal appendix is a controversial issue, but it may be beneficial, especially in children with complex abdominal conditions and difficult intestinal adhesions, taking into account the relatively high lifetime risk of appendicitis. Incidental appendectomy should not be performed in patients who may need this for construction of catheterizable stoma for the bladder or cecum. In order to avoid infectious complications, a normal appendix should not be removed if prosthetic materials are used or a ventriculoperitoneal shunt is encountered during the operation. If ileocecal Crohn's disease is found to be the cause of the symptoms of appendicitis, the appendix should not be removed because there is a significant risk of subsequent fistula formation.

PREOPERATIVE INVESTIGATIONS AND PREPARATION

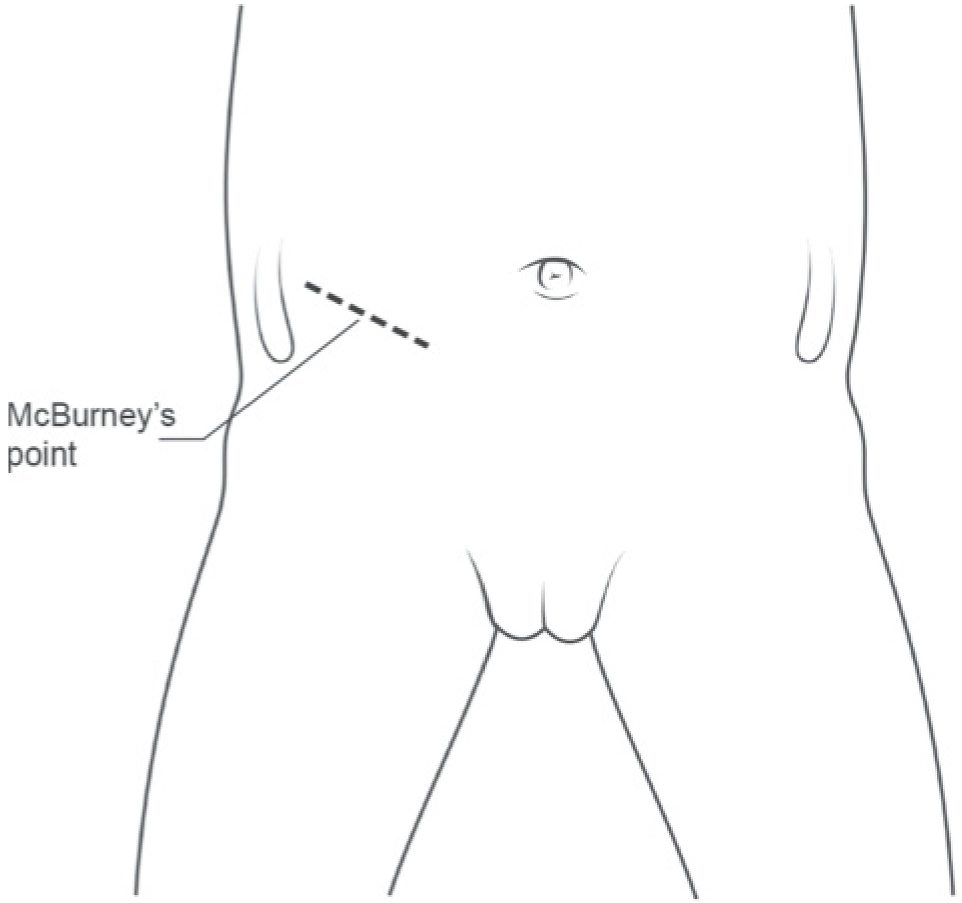
Diagnosis of acute appendicitis still relies on a typical history and clinical findings. In many instances, no further tests are required. Laboratory studies provide limited help in arriving at a definitive diagnosis. Most children will have an elevated left-shifted leukocyte count. Elevated C-reactive protein (CRP) values are helpful for the diagnosis but do not occur in the early phases of the disease. When the diagnosis is questionable, abdominal ultrasonography (US) is useful, not only to visualize an inflamed appendix, but also to exclude other abdominal and pelvic conditions, especially in teenage girls.

Acute appendicitis is usually diagnosed on the basis of the classical clinical symptoms of migratory right iliac fossa pain, nausea, right lower quadrant tenderness, and fever. Children aged over 5 years more often experience a longer delay in diagnosis and higher incidence of perforation. Every effort should be made to confirm the diagnosis of appendicitis before surgery. Formerly reported rates of negative explorations (20–40%) are no longer acceptable. Repeated clinical examinations and appropriate imaging should reduce the negative rates below 10%. Abdominal US is an excellent and accurate screening tool for acute appendicitis. Computed tomography (CT) has slightly higher sensitivity and specificity than US, but the radiation dose remains a significant concern. Therefore, CT with minimized radiation dose is preserved for special situations and complicated patients with an unclear diagnosis and increased operation risk. Significant medical conditions causing appendicitis-like symptoms should be ruled out by clinical examination, laboratory tests, and imaging. These include right basal pneumonia, cholecystitis, meningitis, urinary tract infections, Henoch–Schönlein purpura, and acutely presenting ketoacidosis of type 1 diabetes.

Children with appendicitis should be evaluated for degree of sepsis and dehydration. Most will have vomited or not eaten for more than 24 hours. For this reason alone, preoperative IV fluid resuscitation is mandatory for all patients, with close attention to those with signs of sepsis. Infectious complications are best prevented and treated in all patients by using perioperative antibiotics. Intravenous antibiotic treatment specifically directed against Gram-negative and anaerobic bacteria (e.g. second-generation cephalosporins and metronidazole) is started after the decision of appendectomy has been made. Nasogastric and bladder decompression prior to operation facilitate a laparoscopic procedure. Apart from pain control, premedication is generally unnecessary.

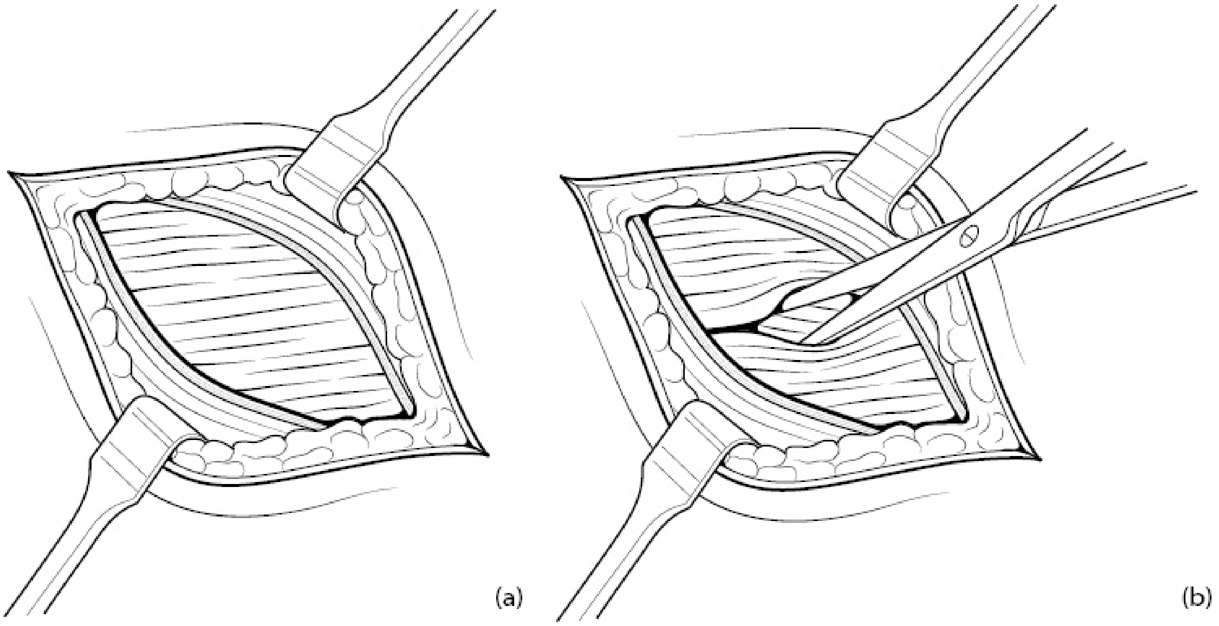
OPEN OPERATION

A transverse or slightly oblique right lower quadrant skin incision is placed slightly above and medial to the anterior superior iliac spine, allowing for extension medially and laterally. The incision courses medially through or below McBurney's point (two-thirds along a line joining the umbilicus with the anterior superior iliac spine, which is the surface marking of the base of appendix) (**Figure 41.1**). Lateral extension of the incision is possible to the flank, while medial enlargement crosses the rectus sheath. In a slender child, the appendix can be removed safely through a 3–4 cm skin incision. As a general rule, a higher and longer incision will provide more room for exploration and dissection but a more prolonged and painful hospital stay.



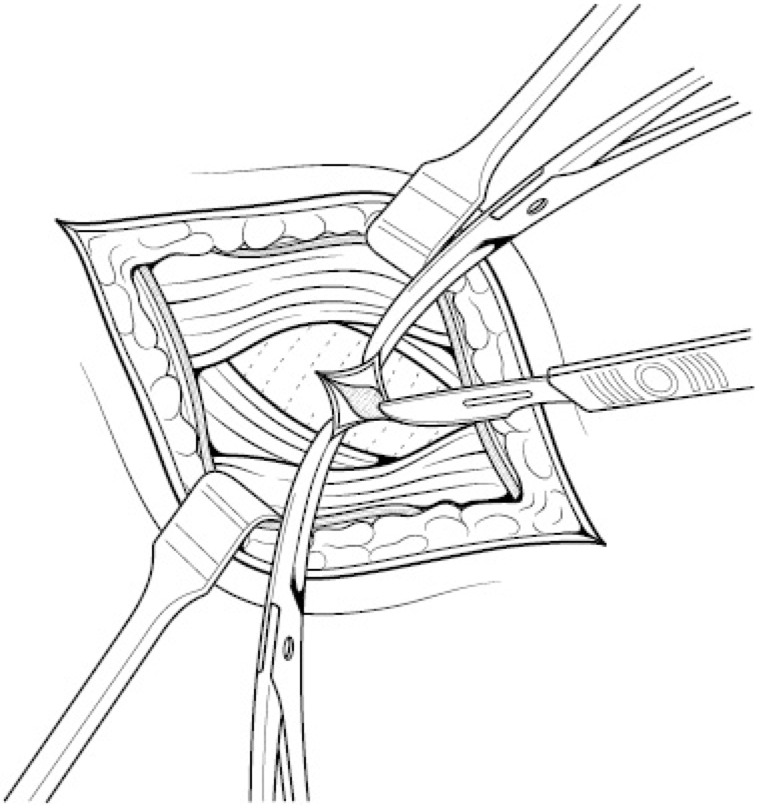
41.1

The external oblique aponeurosis or muscle is split along the direction of the fibers (downward and medial) (**Figure 41.2a**). The internal oblique and transverse muscles are split in the direction of their fibers using blunt-tipped scissors or the fingers of the surgeon (**Figure 41.2b**). Generally, both muscle layers are split in tandem, as the transverse muscle is nearly parallel with the internal oblique muscle. The aponeurosis of the transverse muscle occurs more laterally, and the fibers may run more obliquely from lower right to upper left than those of the internal oblique.



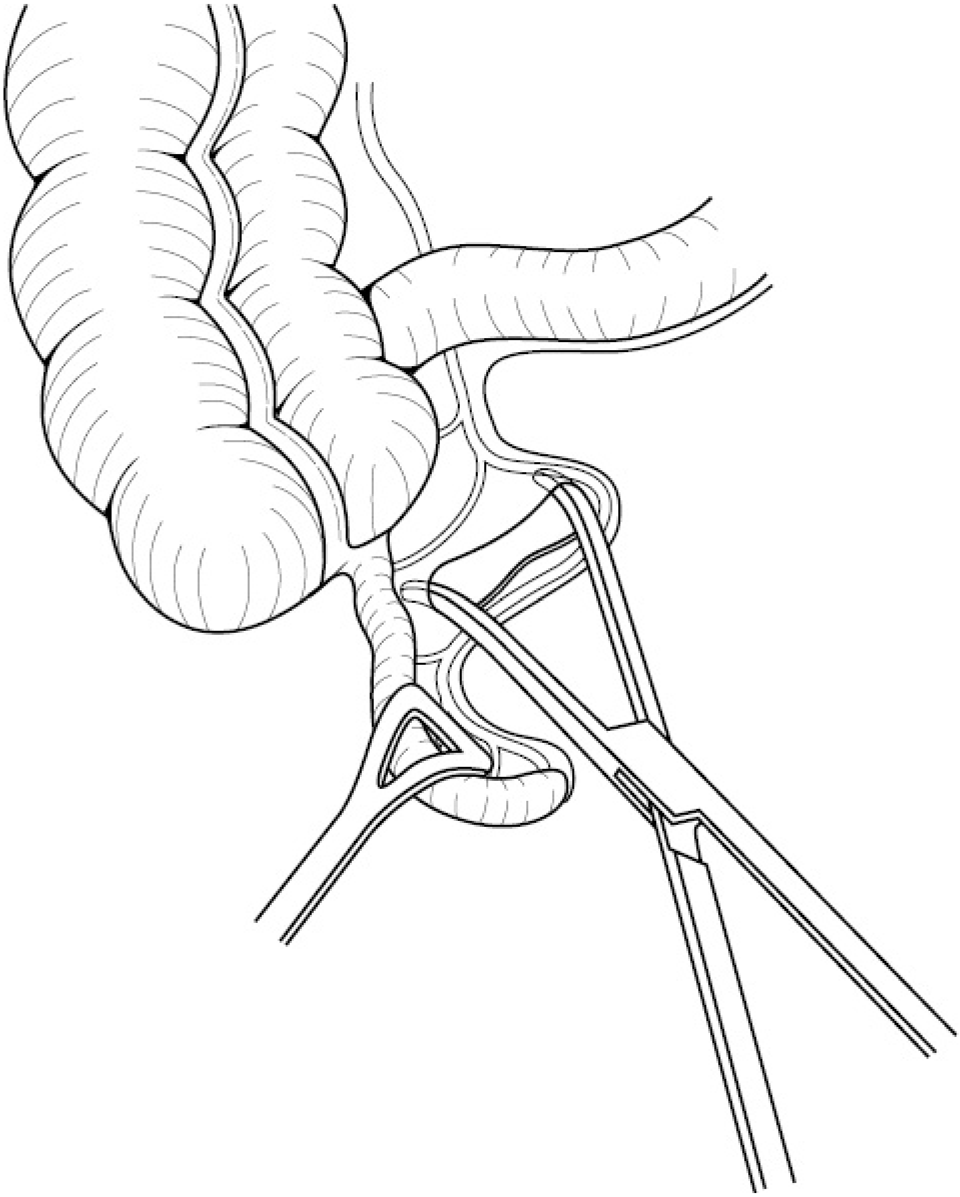
41.2a,b

The peritoneum is grasped after blunt dissection of the transversalis fascia and preperitoneal fat (**Figure 41.3**). Edema from appendiceal inflammation may obscure the peritoneum laterally and it is best identified medially. Free fluid is suctioned on opening the peritoneum with a scalpel and may be sent for culture.



41.3

The key to successful exposure of the appendix is delivery of the cecum into the wound. The anterior teniae coli is grasped and the cecum delivered first pulling it down then up (**Figure 41.4**). If the cecum cannot be delivered easily, lateral peritoneal attachments may require dissection under direct vision using cautery.

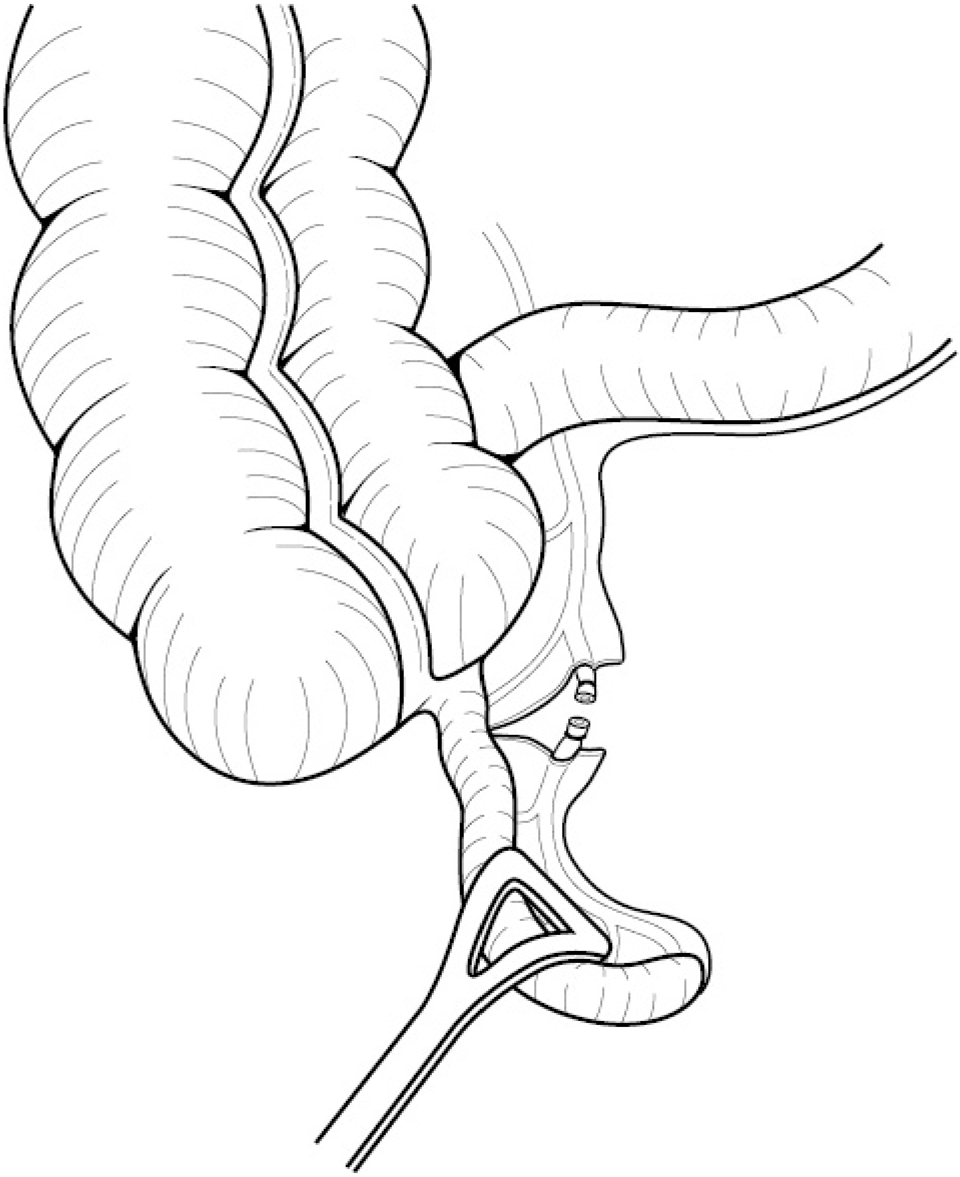


41.4

An appendiceal mass is almost always bound down laterally and inferiorly. Medial attachments are usually to the terminal ileum and its mesentery. In most cases, a large appendix mass cannot be delivered through the wound. However, the appendix itself can usually be mobilized by blunt dissection to enable removal and safe ligation of the appendix base.

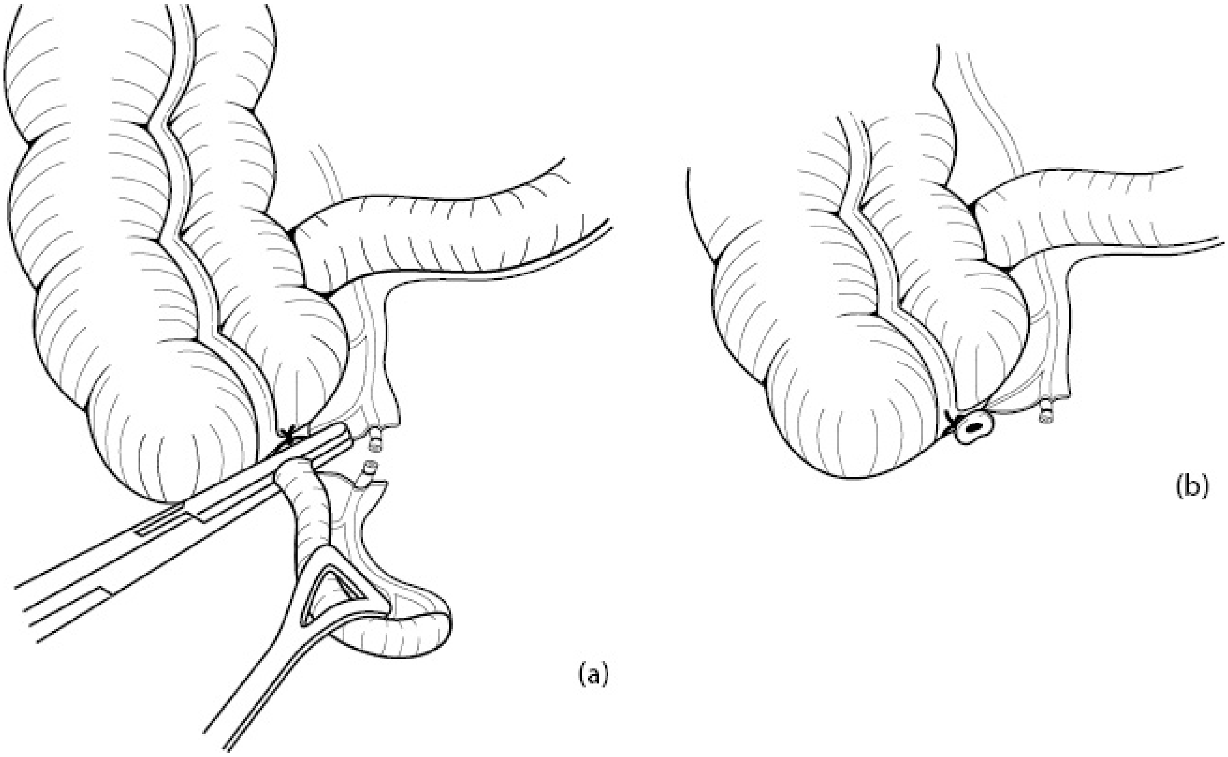
A difficult dissection becomes easier with wound extension. The muscle-splitting incision can be extended medially across the anterior and posterior rectus sheaths using cautery. The rectus muscle is retracted and the inferior epigastric vessels are cauterized or ligated if necessary. Lateral incision enlargement may be helpful if further dissection is required in the flank.

Any attached omentum is divided by cautery or between ligatures, and the mesoappendix is gently grasped with Babcock□ forceps (**Figure 41.5**). With traction on the Babcock□ forceps, the appendix is easily controlled and any further lateral attachments divided with cautery. The mesoappendix is then divided by cautery or between hemostats and ligatures. The mesenteric division should continue to the fold of Treves. Any residual small vessels may be controlled by cautery.



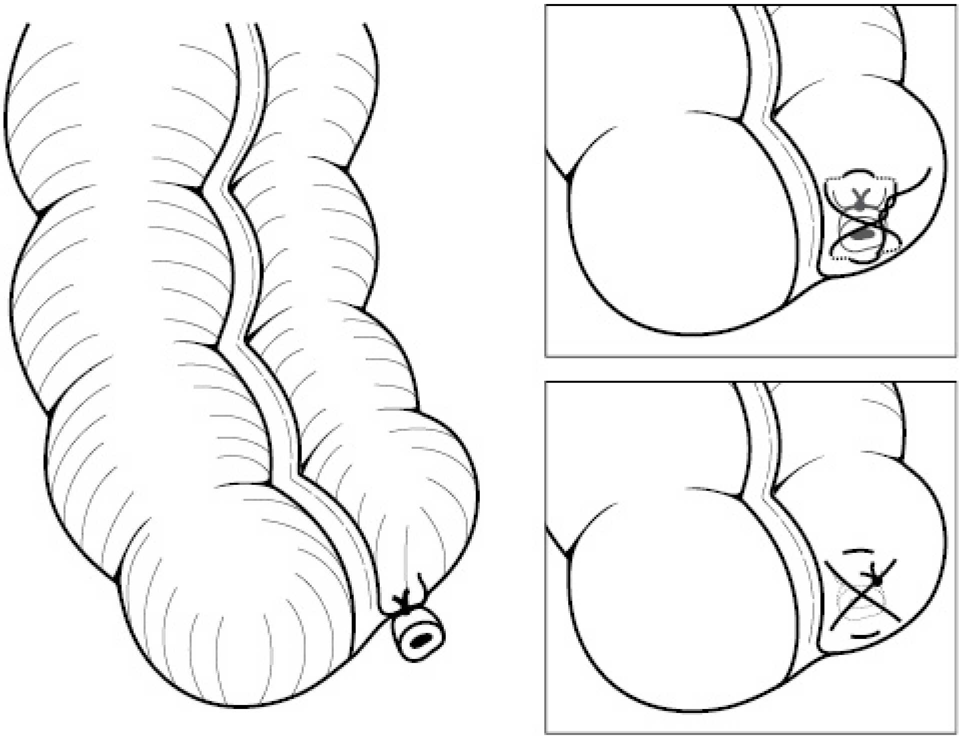
41.5

The base of the appendix is gently crushed just above its origin and the clamp is drawn distally a few more millimeters (**Figure 41.6a**). The appendix is tied at the crushed area and removed by sharp division just proximal to the clamp (**Figure 41.6b**). The mucosa of the remaining stump may be cauterized.



41.6a,b

Stump inversion is not necessary but, if performed, an absorbable Z-stitch or purse-string (aka “tobacco bag”) suture is placed through the seromuscular base of the cecum (**Figure 41.7**). Rarely, the base of the appendix is so severely and widely inflamed that it cannot be safely ligated. In these cases, the whole inflamed base should be inverted by separate stitches that are taken through the healthy cecal wall.



41.7

Before replacing the cecum, the operative field and mesoappendix are inspected for hemostasis. The distended cecum may prove difficult to return to the abdomen. Gentle emptying of the cecum by milking with finger compression usually deflates the bowel to enable its safe repositioning within the abdomen.

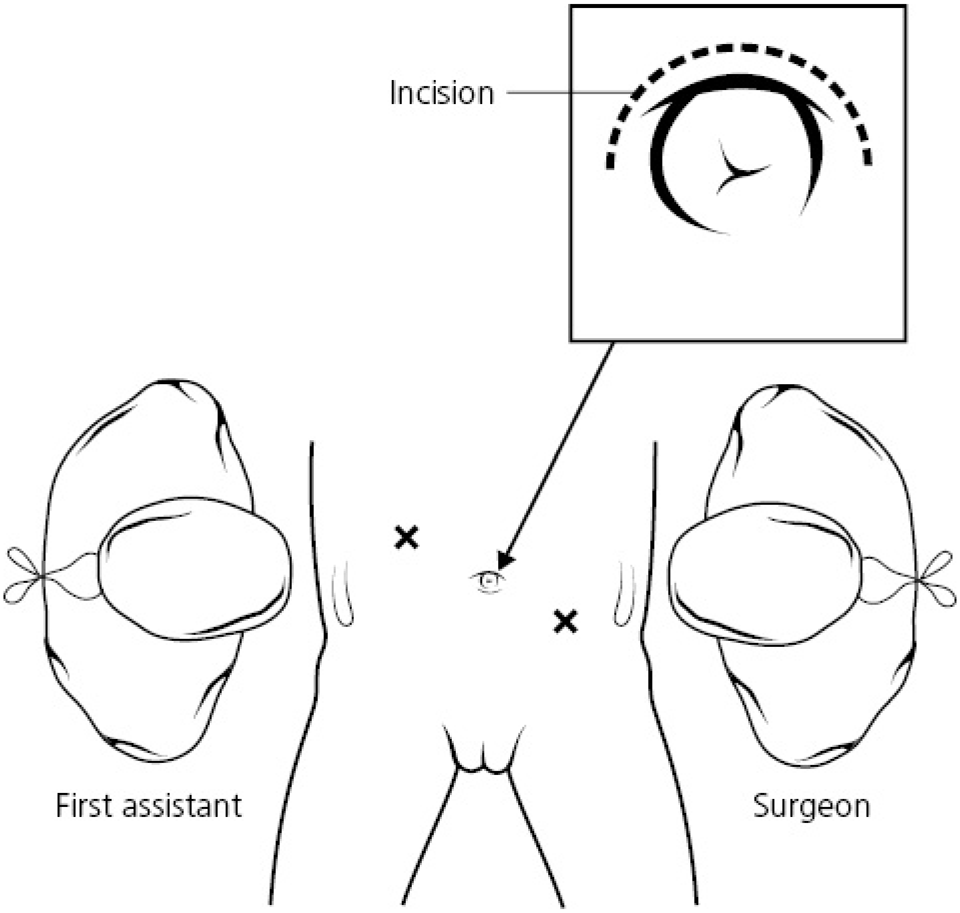
The pelvis and right paracolic spaces are suctioned. In simple appendicitis, wound irrigation is not required. In perforated appendicitis, samples for bacterial culture are obtained and the operative field is irrigated to evacuate all visible pus, but separate wound drainage is not necessary.

The wound is closed in layers with absorbable sutures. The peritoneum is closed with a running suture. The internal oblique and transverse muscles are closed together by apposition of the internal oblique epimysium using interrupted, absorbable sutures. The external oblique aponeurosis is closed with a running suture. Even when there has been perforation and gross contamination, the skin is usually closed with an absorbable, running, subcuticular suture.

If a normal appendix is found, the peritoneal cavity is formally explored for other conditions, such as Meckel's diverticulitis, Crohn's disease, mesenteric lymphadenitis, and gynecological conditions in adolescent females.

LAPAROSCOPIC APPENDECTOMY

The surgeon usually stands on the left side of the patient and a first assistant on the right with personal screens on the opposite sides. A short, 10–15 mm, semicircular incision is made to the upper edge of the umbilicus (**Figure 41.8**). The fascia is exposed and incised transversely. A 5 mm or 12 mm port is inserted under visual control. The abdomen is insufflated with CO_2 using up to 10–12 mmHg intra-abdominal pressure.

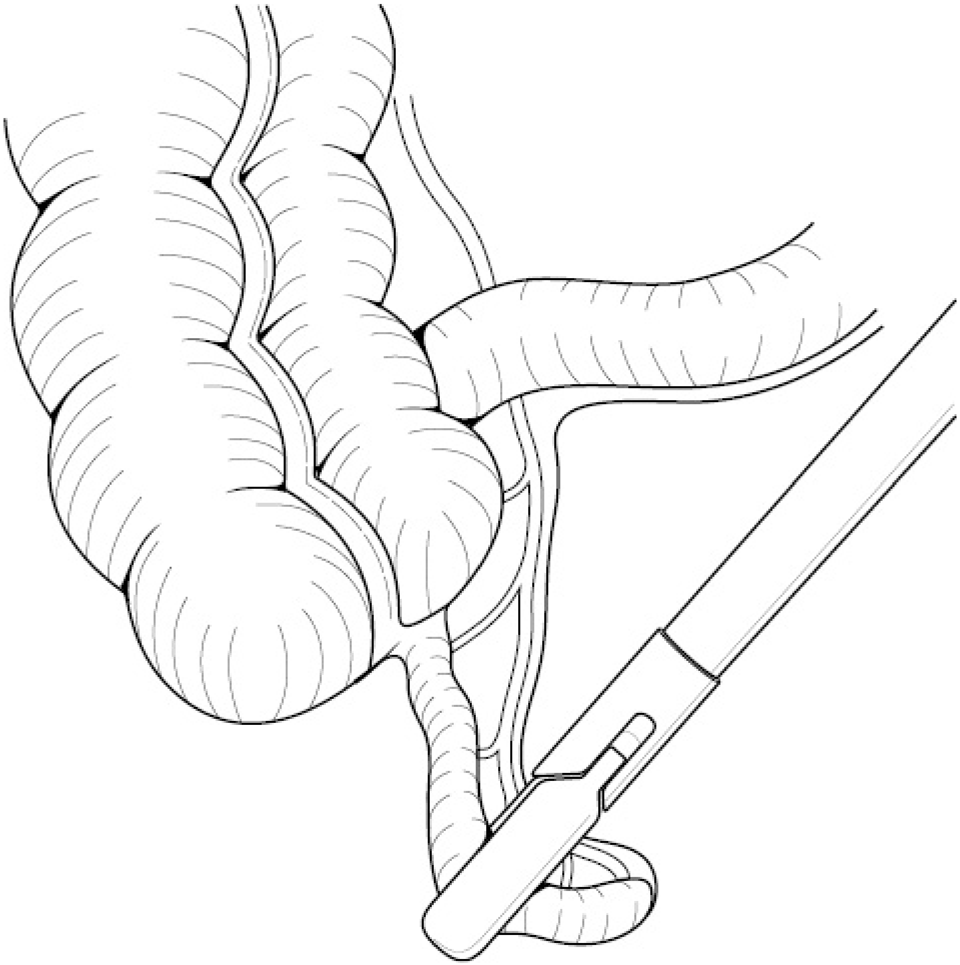


41.8

In addition to the umbilical camera port, two 5 mm working ports are introduced under visual control. One is placed to the right upper quadrant at or above the level of the umbilicus below the liver edge in order to provide enough working space; the other is placed to the left lower quadrant to the level of the iliac spine. Alternative port placements substitute the right upper quadrant one for a midline suprapubic one.

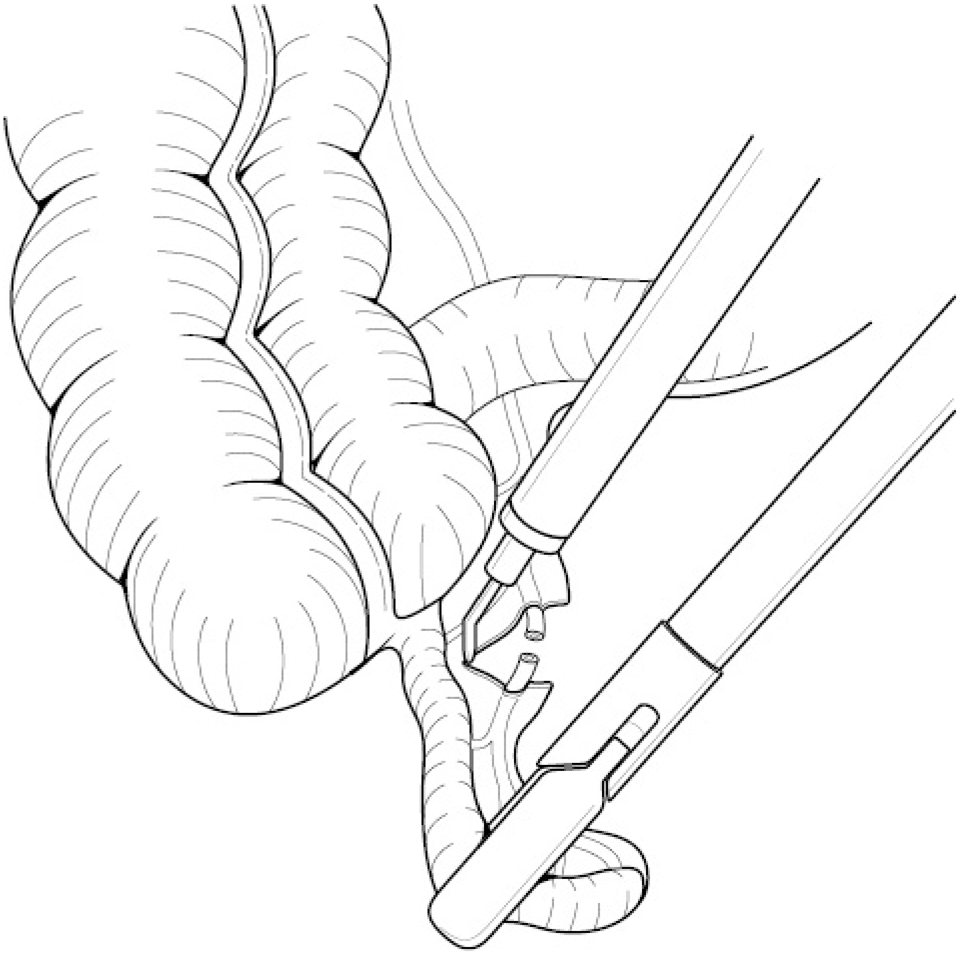
After the laparoscopic ports have been introduced, the patient is turned to a generous Trendelenburg position and rotated to the left for optimal laparoscopic exposure of the appendix.

The distal mesentery of the appendix is grasped by Maryland or Babcock□ forceps to expose the base of the appendix (**Figure 41.9**). In a case of an uninflamed appendix, the entire abdominal cavity is carefully assessed including the entire length of the small intestine from the ileocecal valve to the ligament of Treitz and ovaries in girls.



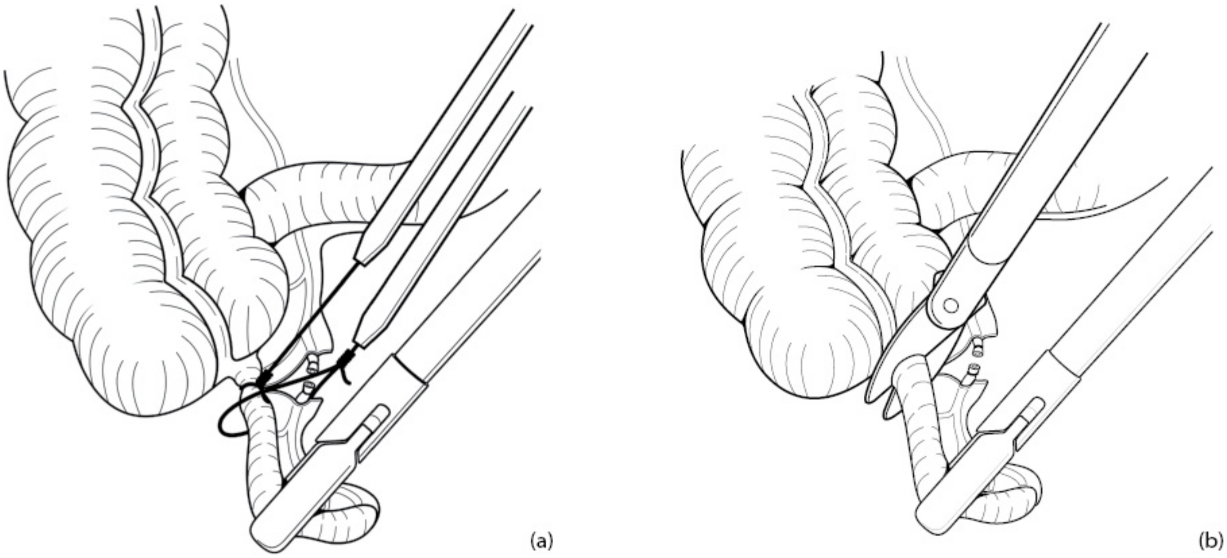
41.9

The appendix is mobilized by dividing inflammatory adhesions to the surrounding structures, such as the omentum or a loop of small bowel. Division is accomplished with a diathermy hook or using blunt dissection (**Figure 41.10**). In the case of a retrocecal appendix, the cecum is mobilized first according to the same principles. The mesentery and its vessels are divided at the level of the base of the appendix using a diathermy hook.



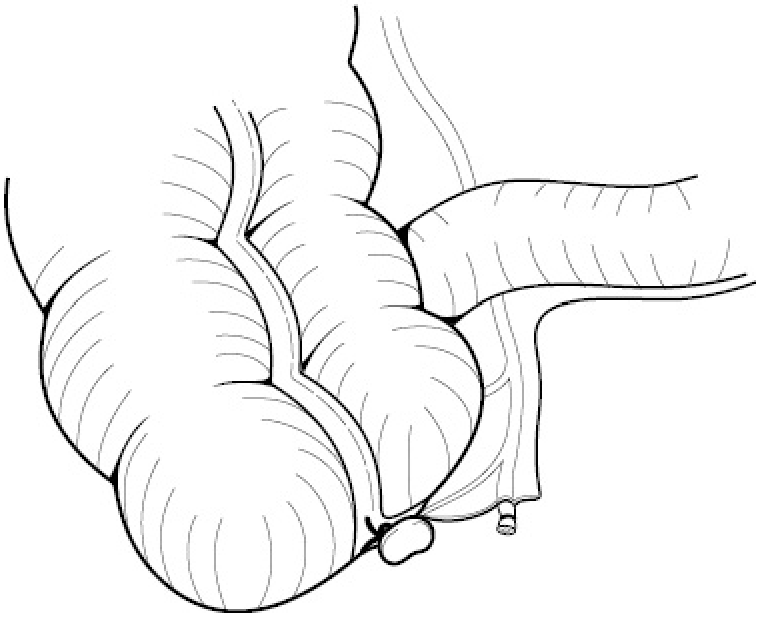
41.10

The base of the appendix is ligated twice with a laparoscopic pre-tied loop of absorbable material (e.g. ENDOLOOP□) (**Figure 41.11a**). The appendix is then grasped distally and divided with scissors (**Figure 41.11b**). Rarely, when the entire appendix including the base is necrotic, it is safer to use an endoscopic cutting stapler device by including the healthier tissue of the cecum in the staple line. Inversion of the appendiceal stump is not necessary.



41.11a,b

An inflamed appendix may be removed through a trocar or using a laparoscopic bag, avoiding direct contact with port incisions. Visible pus is removed with a combination of irrigation and suction before the cavity is lavaged with liberal quantities of saline. The operative field is checked for hemostasis, the abdomen is desufflated, and trocars are removed. Only the umbilical incision is closed with fascial sutures (**Figure 41.12**). The skin incisions are closed with intracutaneous sutures or simply glued.



41.12

POSTOPERATIVE CARE

Postoperative nasogastric decompression is indicated only in cases with extensive intra-abdominal contamination or obvious associated small intestinal obstruction due to appendicitis. The need for abdominal drainage is exceptional. In non-perforated cases, the patients are allowed to resume oral intake once recovered from the anesthesia and the IV antibiotics are discontinued after three doses. Children with uncomplicated appendectomy are usually discharged within 2 days after both open and laparoscopic procedures. Following perforated or gangrenous appendicitis, IV antibiotics are switched to oral ones when the temperature has normalized and/or CRP levels are clearly decreasing. Oral antibiotics are continued until CRP has normalized. Oral food intake is restarted along with resolution of ileus and gradual reduction of IV fluids. Anti-inflammatory analgesics are usually sufficient for postoperative pain medication.

COMPLICATIONS AND OUTCOMES

Surgical complications are more common after complicated appendicitis and include wound infection, intra-abdominal abscess formation, hemorrhage, prolonged paralytic ileus, and intestinal obstruction. Appendix stump leak is extremely uncommon. Surgical site infection (SSI) is by far the most common complication but with achievable rates of <5% even for complicated appendicitis. SSI can be successfully managed with local drainage, wound care, and oral antibiotics. Frequency of postoperative intra-abdominal abscess formation is around 2% and should be confined to those who have had a perforated appendix. Location and extent of postoperative abscesses are best assessed with CT. Treatment includes systemic antimicrobial therapy with or without percutaneous US-guided drainage. Intestinal obstruction following appendectomy occurs in <1% of children with appendicitis, but it may necessitate early reoperation and adhesiolysis, especially among preschool-aged children.

VIDEO

OPS_8e_video_41.1 Laparoscopic appendectomy

FURTHER READING

Almström M, Svensson JF, Patkova B, et al. In-hospital surgical delay does not increase the risk for perforated appendicitis in children: A single-center retrospective cohort study. *Ann Surg* 2017; 265: 616–21.

Hall NJ, Eaton S, Stanton MP, et al. Active observation versus interval appendicectomy after successful non-operative treatment of an appendix mass in children (CHINA study): An open-label, randomised controlled trial. *Lancet Gastroenterol Hepatol* 2017; 2: 253–60.

Svensson JF, Patkova B, Almström M, et al. Nonoperative treatment with antibiotics versus surgery for acute nonperforated appendicitis in children: A pilot randomized controlled trial. *Ann Surg* 2015; 261: 67–71.

van Heurn LWE, Pakarinen MP, Wester T. Contemporary management of abdominal surgical emergencies in infants and children. *Br J Surg* 2014; 101: e24–e33.

42

Necrotizing enterocolitis

NIGEL J. HALL

HISTORY

Necrotizing enterocolitis (NEC) is a devastating disease of infancy and the most common serious acquired gastrointestinal (GI) disease in the newborn infant. The term "necrotizing enterocolitis" was first coined in the 1950s when used to describe infants who died with necrotic lesions of the GI tract, but it did not become recognized as a distinct clinical entity until the 1960s, when a number of authors began reporting their experience with this disease. With improvements in neonatal intensive care over the past four decades, the incidence of NEC is increasing as more babies born at the extreme limits of prematurity now survive. Recent estimates place the incidence at about 0.5% of all live births and at about 5% of very low birth weight infants. Concurrent with this increase in incidence has been an extensive amount of time and energy devoted to exploring the etiology and pathogenesis of this disease by a number of individuals and groups worldwide. Despite these efforts, our understanding of the processes contributing to the development of NEC remains limited and, as a result, we are unable to provide specific focused treatments.

PRINCIPLES AND JUSTIFICATION

NEC is a disease state comprising a wide spectrum of intestinal pathology and resulting systemic manifestations. At one end of the spectrum lie infants with minimal systemic upset with a distended abdomen and in whom the diagnosis of NEC is made on the pathognomonic radiographic finding of pneumatosis intestinalis or portal venous gas. In the absence of evidence of intestinal perforation or clinical deterioration, such infants are often managed conservatively by resting and decompressing the intestinal tract and administering broad-spectrum antibiotics. Following 7–10 days of such conservative management, feeds may be slowly reintroduced. While some of these infants may develop complications of NEC such as a post-inflammatory intestinal stricture requiring surgery, many recover without the need for operative intervention.

At the other end of the spectrum are infants with extensive intestinal involvement and gross systemic upset, often with failure of multiple organ systems. These infants almost invariably require surgical intervention and often require resection of one or more lengths of gangrenous intestine. Unfortunately, in these severely ill babies the intestinal involvement is only part of the overall disease process, and the development of multisystem organ failure requires prolonged periods of intensive care. Mortality in this severely affected group maybe as high as 50%.

There is a group of infants who present with similar, if not identical, abdominal and systemic symptoms and signs to those found in infants with NEC, but who are found to have a focal or spontaneous intestinal perforation (SIP), often in the terminal ileum, with preservation of the remainder of the GI tract. This is often associated with non-steroidal anti-inflammatory therapy used to encourage closure of a patent ductus arteriosus and is seen particularly in extremely low birth weight infants. While SIP has been considered one end of the spectrum of NEC by some, there is now emerging consensus that this is a different disease entity, with different etiology, pathogenesis, and clinical presentation from NEC. However, from the perspective of an operative surgery textbook, surgical procedures performed are similar to those utilized for NEC so it remains appropriate for SIP to be mentioned in this chapter.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

The onset of symptoms of NEC appears to be inversely related to the gestational age at birth and birth weight, such that full-term infants who develop NEC (primarily but not exclusively those with congenital cardiac abnormalities) do so in the first few days of life, whereas preterm infants of low birth weight develop symptoms most commonly between 15 and 20 days of age. The etiological factors implicated in the development of NEC are an immature intestine, mesenteric hypoxia or ischemia, bacterial colonization of the intestine, and the presence of feed within the intestinal lumen. The author notes that frequently there has been a change in feed type in an infant who develops NEC within the preceding 7 days. The only factor recognized to reduce the incidence of NEC is feeding with maternal expressed breast milk.

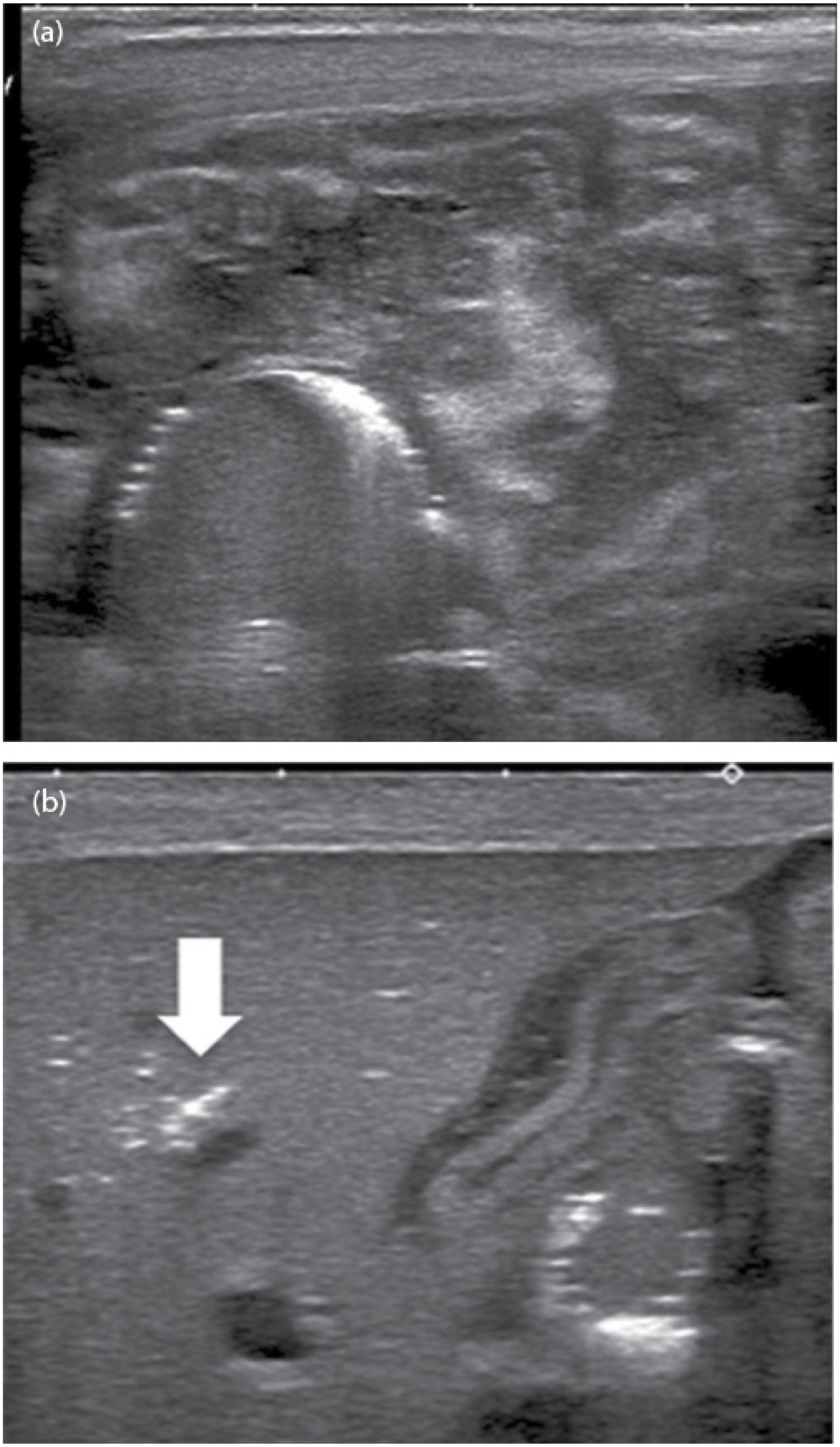
A significant challenge in the care of an infant with NEC is distinguishing those infants who will recover with medical treatment alone from those who will require surgical intervention. On occasion, the initial presentation is that of an acutely unwell infant with a systemic inflammatory response, multi-organ failure, grossly distended abdomen, and evidence of GI perforation on abdominal radiograph. In such cases, few would argue against surgical intervention. However, more frequently, an infant with NEC presents with a less severe illness and sometimes with very subtle clinical features. In these infants, the need for surgical intervention is less clear-cut and such infants are usually monitored for a short period of time (e.g. 24–48 hours) to determine how their illness progresses before deciding on the need for surgery. This will typically involve serial clinical observation and usually some form of repeated radiological imaging. Close liaison between neonatal and surgical teams is recommended in these cases to aid appropriate decision making.

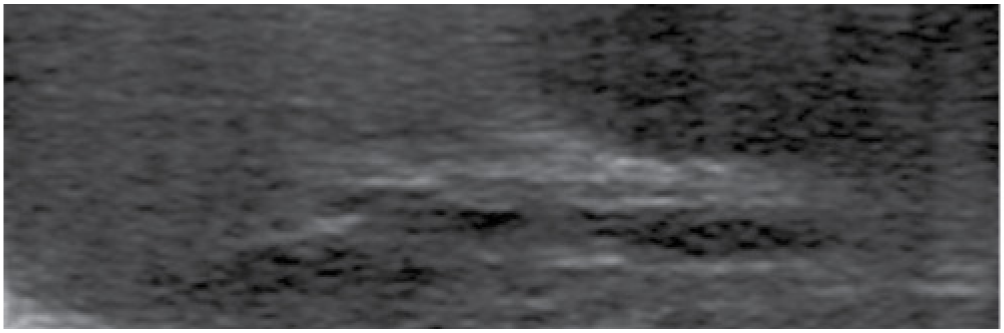
Whether being treated initially non-operatively or surgically, adequate resuscitation is essential, with appropriate attention and interventions to support respiratory and cardiovascular systems in particular. Empirical broad-spectrum antibiotic therapy, gastric decompression, and cessation of enteral feeds are standard non-operative treatment from the point of diagnosis. Particular attention should be made to monitor for thrombocytopenia and coagulopathy and to treat at appropriate thresholds. This is particularly important in infants being prepared for surgery.

Radiographic features

The radiographic findings in infants with NEC vary from one case to another, but the presence of pneumatosis intestinalis is pathognomonic of NEC. The other finding highly suggestive of NEC even in isolation is that of "portal venous gas" seen both radiologically and on an abdominal ultrasound (US) scan. A number of radiological signs may indicate the presence of a pneumoperitoneum secondary to an intestinal perforation (seen in both NEC and SIP) although signs of pneumoperitoneum can be subtle. Serial imaging is recommended if a conservative strategy is adopted. The surgeon is essentially looking for radiological signs that may support the need for surgery as opposed to ongoing non-operative treatment. For example, a newly appearing pneumoperitoneum or a "fixed intestinal loop" (i.e. dilated loops of intestine which appears identical in size and location on serial radiographs). The latter may be indicative of intestinal ischemia or necrosis and is taken as an indication for surgery by some surgeons. Of note, radiographs of infants with SIP typically have none of these findings other than pneumoperitoneum.

While plain radiography has long been the mainstay of radiological investigation for infants with NEC, abdominal US has recently become more commonplace. US is a highly sensitive modality for the visualization of pneumatosis intestinalis (Figure 42.1a), portal venous gas (Figure 42.1b) and free intraperitoneal gas. Furthermore, US permits evaluation of the intestinal wall, peristalsis and intestinal blood flow. Key sonographic findings have been proposed to be indicative of intestinal ischemia and necrosis and the author finds value in these when evaluating infants for surgical intervention. Indications for surgical intervention in the setting of acute NEC are shown in Table 42.1.


(a)
(b)



42.1 (a) Ultrasound image showing multiple foci of intramural gas on transverse view of bowel loop. (b) Ultrasound image showing portal venous gas (arrow).

Table 42.1 Indications for surgery in infants with acute NEC

Absolute	Relative
Pneumoperitoneum	Increased abdominal tenderness, distension, and/or discoloration
Clinical deterioration despite maximal medical treatment	Fixed intestinal loop
Abdominal mass with persistent intestinal obstruction or sepsis	US suggesting intestinal necrosis
	Portal vein gas
	Thrombocytopenia

In addition to surgery during the acute stage of NEC, a number of infants require surgery at a later stage to treat complications of NEC regardless of whether they have had surgery during the acute stage of their illness. The most common indication is the presence of an intestinal stricture.

Surgical approach

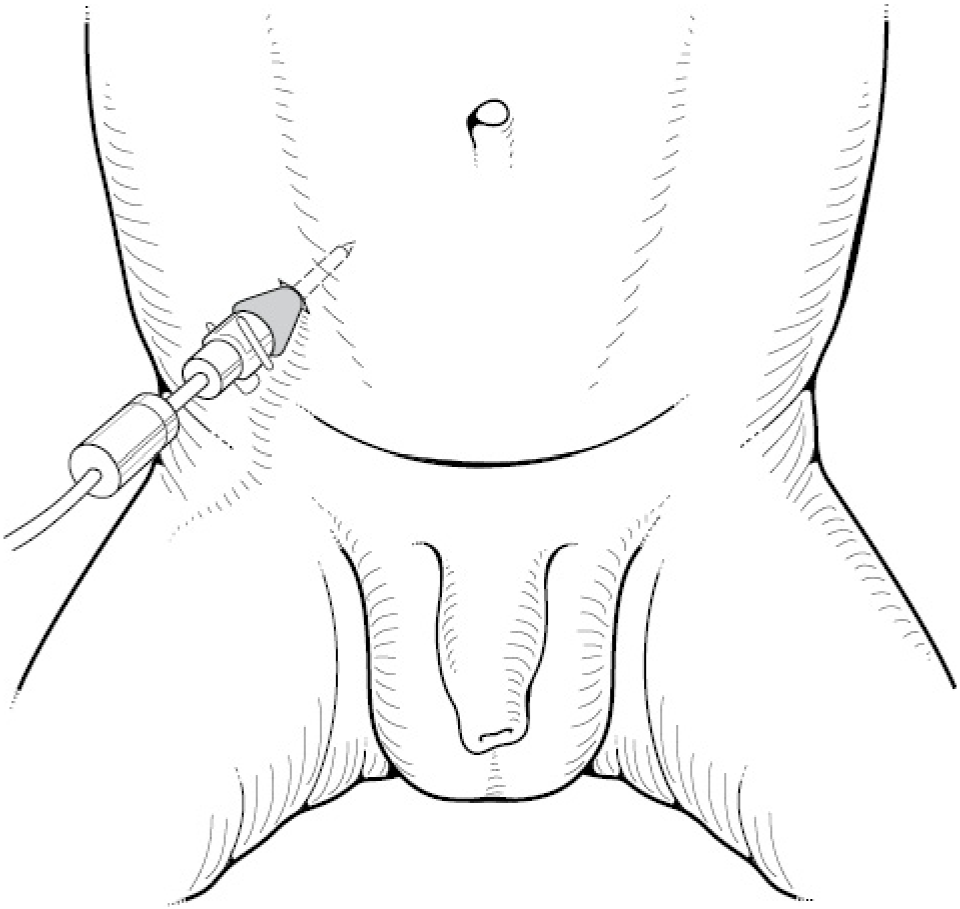
There is little surgical consensus among surgeons concerning the ideal surgical management of NEC. Surgical options include primary peritoneal drainage (PPD) or laparotomy. PPD was first reported in 1975 by Marsh and Ein from Toronto as a means of stabilizing and improving the systemic status of premature infants with intestinal perforation secondary to NEC. Since then, PPD has been reported as a definitive therapy in infants with pneumoperitoneum rather than an intermediary for laparotomy. Two large multicenter randomized controlled trials have demonstrated no benefit of PPD over laparotomy even in very small infants (<1000 g). The author's practice for infants with NEC (as opposed to SIP) is to reserve PPD as a decompressive temporary measure while stabilizing the infant for laparotomy.

Minimal access surgery has become commonplace in recent years within the field of pediatric surgery but its incorporation into the mainstream surgical treatment of infants with NEC has not been successful. The principal limitation of minimal access surgery as definitive treatment for NEC is physical size of the child. However, diagnostic laparoscopy may assist surgical decision making in infants with NEC, or in an unwell infant with an acute abdomen where NEC is one of the differential diagnoses. Laparoscopy may be used to provide visualization of the intestine and therefore a more informed decision to be made concerning the need for laparotomy based on the condition of the intestine.

Diagnostic laparoscopy

Laparoscopy may be performed on the neonatal intensive care unit under fentanyl anesthesia with muscle paralysis. High-frequency oscillatory ventilation and inhaled nitric oxide therapy do not preclude laparoscopy.

A 5 mm Hasson cannula is inserted in the right or left iliac fossa under direct vision and CO_2 pneumoperitoneum is established with a maximum pressure of 10 mmHg and maximal flow rate of 2 L/min (Figure 42.2). A 30-degree laparoscope is inserted and the intestine and intraperitoneal contents are inspected.



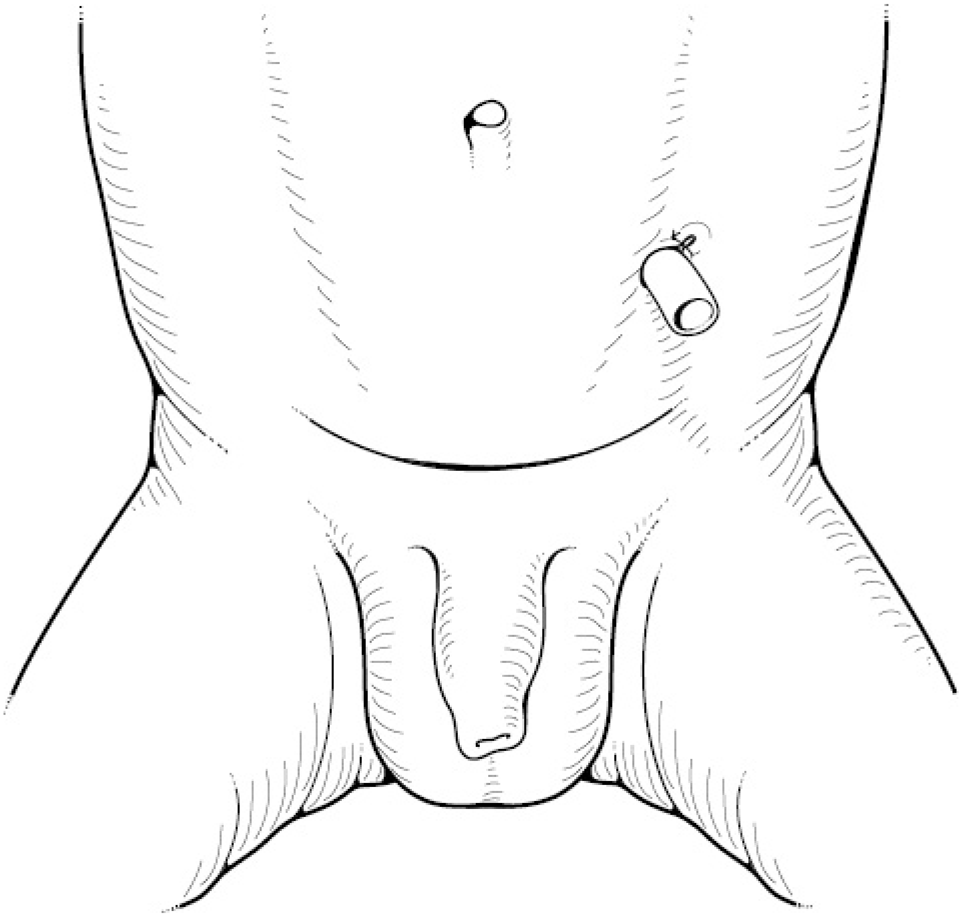
42.2

In cases in which there is perforation with peritoneal contamination, an additional incision may be made in the abdominal wall for the introduction of a peritoneal suction/irrigation device. Identification of frankly gangrenous intestine during laparoscopy may be considered an indication for laparotomy.

PRIMARY PERITONEAL DRAINAGE

PPD can be performed at the cot-side with the infant on the neonatal intensive care unit. The site of drain insertion is usually the left iliac fossa, although the right iliac fossa may also be considered if the lower border of the liver can clearly be palpated above the insertion site. The upper quadrants of the abdomen should be avoided to limit visceral damage.

Local anesthetic is infiltrated into the skin and subcutaneous tissues. A small (0.5–1 cm) incision is made in the skin and blunt dissection used to expose first the fascia and then the peritoneum, which is carefully opened to avoid damage to the underlying bowel. Gas or meconium-stained fluid may be released from the peritoneal cavity and a microbiological swab should be taken to direct future antibiotic therapy. A soft drain (e.g. Penrose drain or alternatively a finger from a surgical glove) is inserted into the peritoneum and sutured in place to the skin to allow continued drainage of the peritoneal cavity (Figure 42.3). Stiff drains, including IV cannulas, are best avoided as they may perforate the intestine or cause visceral damage.



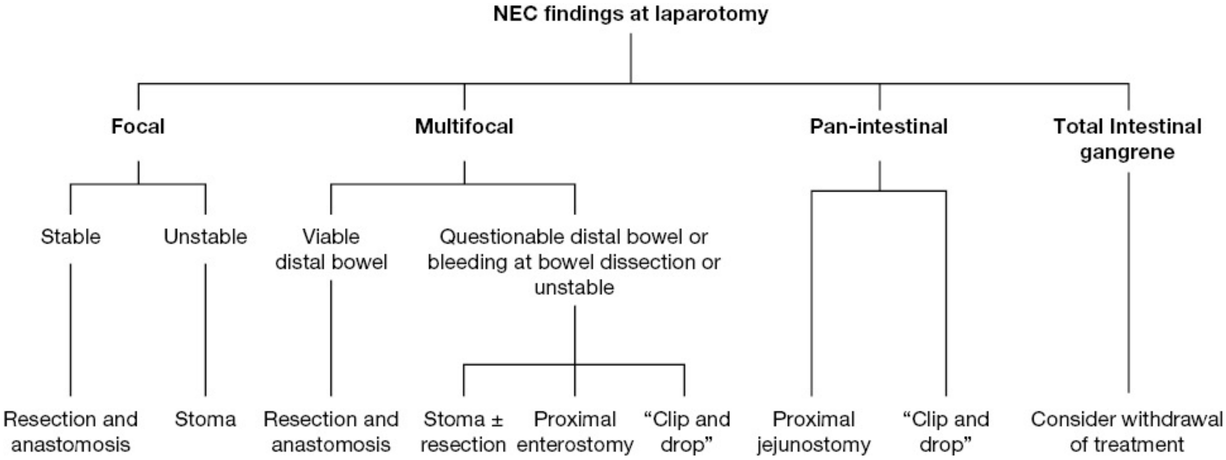
42.3

LAPAROTOMY

The principal surgical objectives of laparotomy in acute NEC are to control sepsis, remove gangrenous bowel, and preserve as much bowel length as possible. Within these objectives, a number of options exist. The patient's weight and clinical status, as well as the extent of the disease, influence the choice of surgical procedure. At laparotomy, the extent of the disease can be classified as "*focal*" when it is limited to a single intestinal segment, "*multifocal*" if it includes two or more intestinal segments with >50% of the small intestine viable, and "*pan-intestinal*" when the majority of small and large bowel is involved with <25% viable bowel remaining.

Laparotomy is performed under general anesthesia and may be performed on the neonatal intensive care unit or in the operating room, depending on the stability of the infant and local practice and policy.

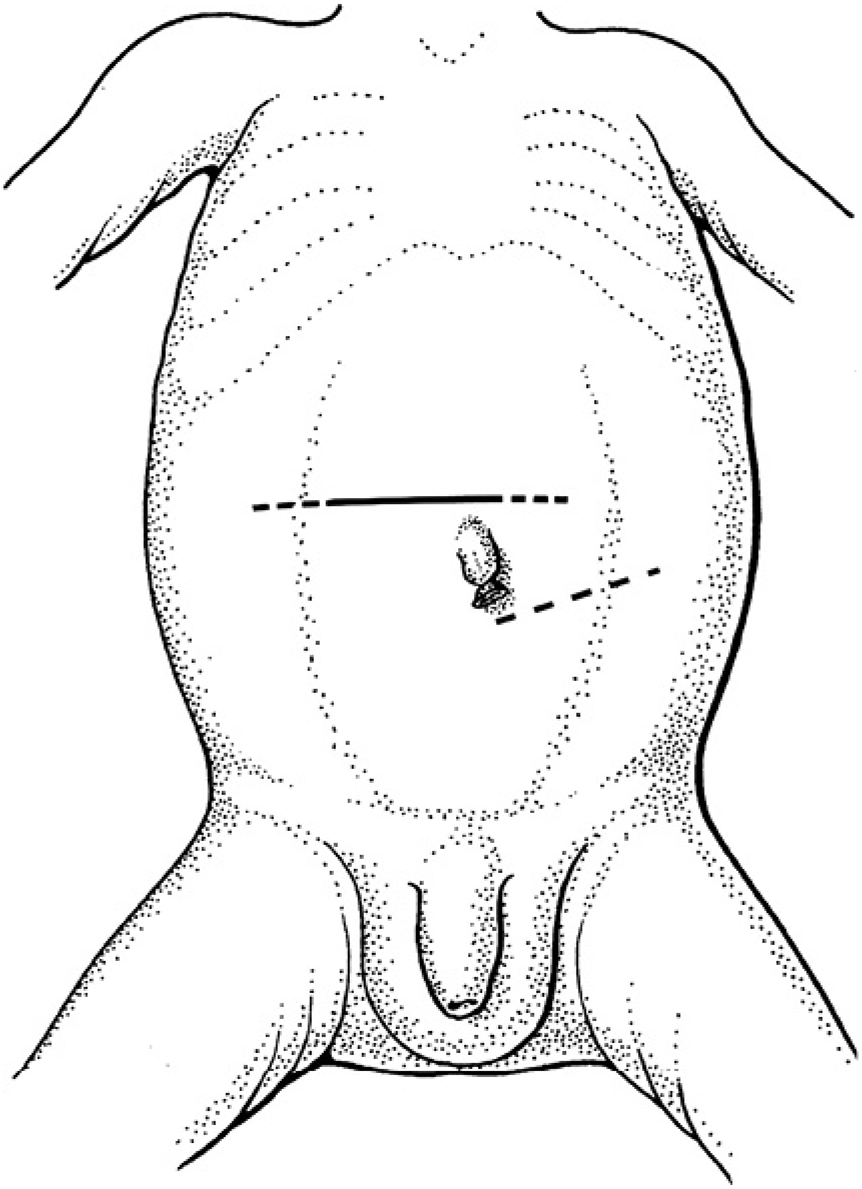
The author's preferred approach to the operative management at laparotomy of infants with NEC is illustrated in Figure 42.4.


NEC findings at laparotomy
Focal
Multifocal
Pan-intestinal
Total Intestinal gangrene
Stable
Unstable
Viable distal bowel
Questionable distal bowel or bleeding at bowel dissection or unstable
Resection and anastomosis
Stoma
Resection and anastomosis
Stoma ± resection
Proximal enterostomy
"Clip and drop"
Proximal jejunostomy
"Clip and drop"
Consider withdrawal of treatment

42.4

Incision

A transverse supraumbilical incision is used for laparotomy. This may be slightly to the right side initially, allowing for further extension of the incision to the left side if necessary (Figure 42.5). In infants <1000 g, an oblique infraumbilical incision on the left side of the abdomen may be preferred since this still permits access to the entire abdominal cavity but moves the incision away from the edge of the vulnerable liver.



42.5

Care should be taken to limit damage to the thin anterior abdominal wall in extremely premature infants. The skin is incised and the underlying subcutaneous tissues and muscle can be safely divided with point diathermy, which has the added advantage of providing a degree of hemostasis. When entering the peritoneal cavity, particular attention should be given to the liver, which is often very large and extremely fragile in infants with NEC. Capsular liver damage should be avoided at all costs, as hemorrhage from any injury to the liver may have disastrous consequences and result in death.

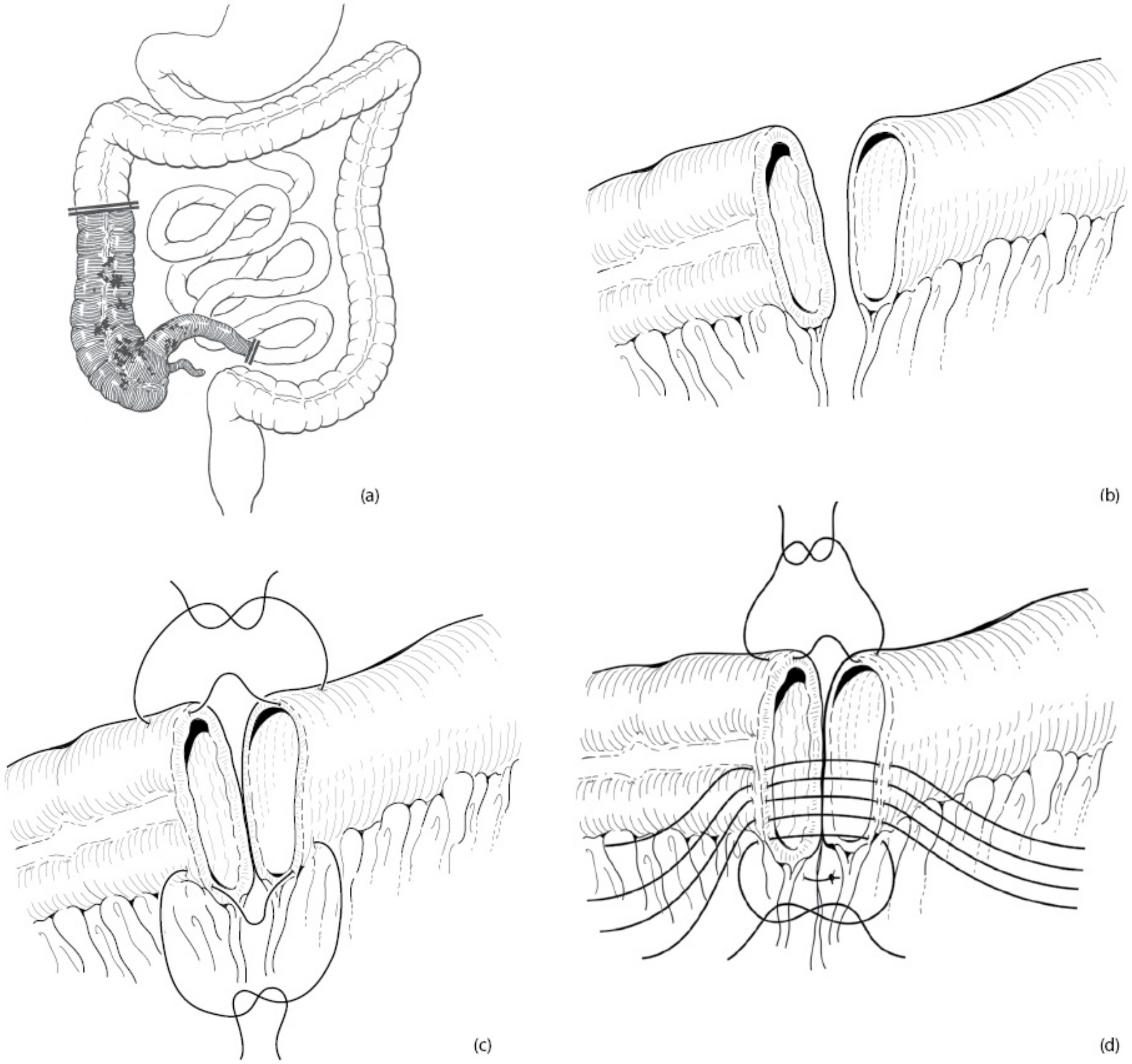
Operations for focal necrotizing enterocolitis

For infants with focal NEC, whether perforated or not, and for infants with a focal intestinal perforation with no evidence of NEC, there are two surgical options at laparotomy, namely resection followed by primary anastomosis and resection followed by stoma formation. Both are valid options and there is no good evidence to support one over the other. While resection followed by primary anastomosis has the advantage of restoring intestinal continuity in one operation and avoids the potential complications associated with stoma formation, there is a risk of anastomotic leakage or stenosis with primary anastomosis. Infants selected for primary anastomosis should be stable during the perioperative period, and the resection margins and remaining (distal) intestine should be healthy with good perfusion. Neither the weight of the child nor the presence of peritoneal contamination secondary to intestinal perforation affects the decision of whether to perform primary anastomosis or to fashion a stoma. Infants who do not fulfill the above criteria should be considered candidates for stoma formation. Although a stoma diverts the fecal stream and avoids the potential complications of primary anastomosis, stoma-related complications in this population are frequent, including excessive fluid and electrolyte losses, prolapse, skin excoriation, and poor growth. The author's preferred approach is to perform a primary anastomosis whenever appropriate while acknowledging that many surgeons feel more comfortable forming a stoma.

RESECTION WITH PRIMARY ANASTOMOSIS

The standard laparotomy incision is used. The affected segment of intestine is delivered through the wound and placed on povidone–iodine-soaked gauze to minimize the risk of further peritoneal contamination.

The affected segment along with its associated mesentery is resected, ensuring hemostasis of the mesenteric vessels, which can be satisfactorily achieved using bipolar diathermy (Figure 42.6a). A single-layer extramucosal, seromuscular anastomosis is performed using an appropriately sized (6/0 in the premature neonate) interrupted monofilament suture (**Figure 42.6b**). The first two stitches are placed on opposite sides of the intestine to give stability (**Figure 42.6c**). The anastomosis is then completed, laying the knots on the outside of the lumen (**Figure 42.6d**). Finally, the mesenteric defect is closed. In instances in which there is a discrepancy in the circumference of the two ends to be anastomosed, this technique may still be used, adjusting the distance between adjacent sutures on one side of the anastomosis. Discrepancies of up to 6 : 1 or 7 : 1 can be accommodated in this way. Alternatively, some surgeons find it helpful to cut one of the ends at an angle to increase the circumference for anastomosis.



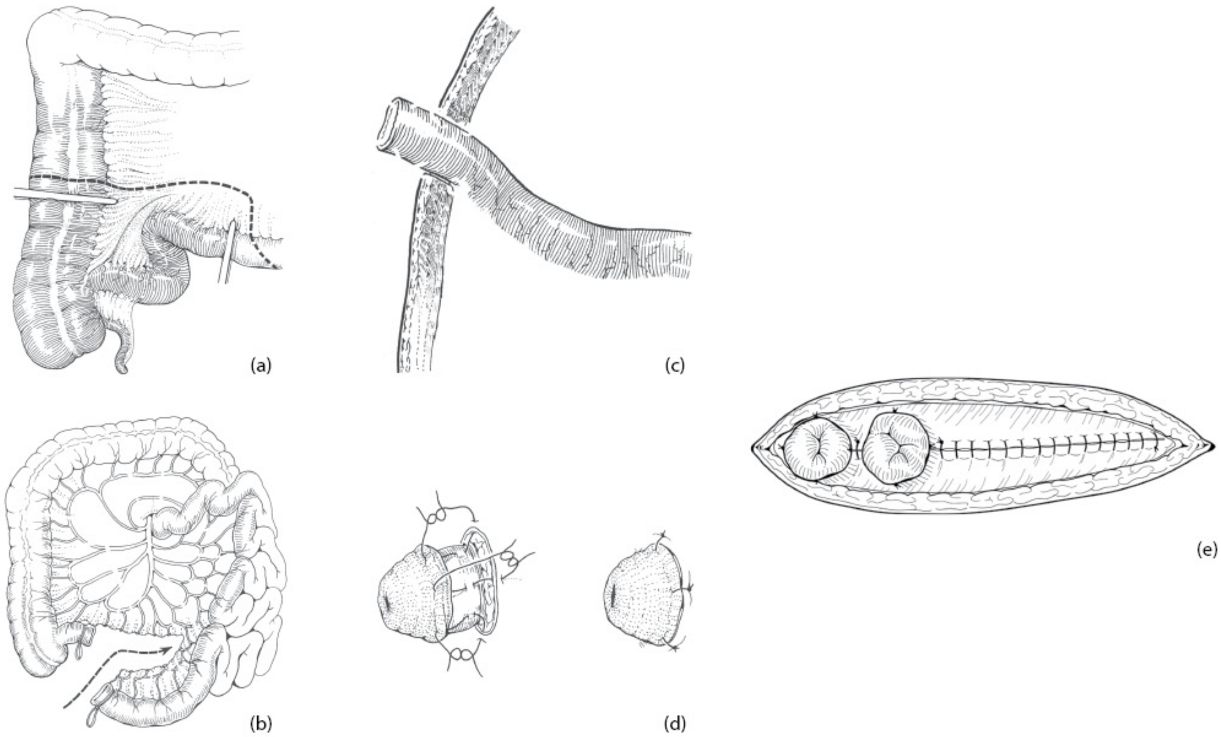
42.6a–d

Providing an adequate length of intestine remains, primary anastomosis may be used following resection of any part of the intestinal tract from the jejunum to the sigmoid colon, including cases of total colonic NEC requiring total colectomy and ileorectal anastomosis.

RESECTION AND STOMA FORMATION

Although a stoma may be fashioned into one end of the abdominal incision, the author's preference is to site the stoma away from the incision if at all possible to reduce the risk of wound and stomal complications

(Figure 42.7a–e). Often, the site of the stoma is determined by the mobility of the mesentery. Whenever possible, a mucous fistula is also fashioned next to the proximal stoma to prevent the need for full laparotomy rather than a more limited procedure when restoring intestinal continuity. When it is not possible to bring the distal resection margin to the abdominal wall, it may be clipped or oversewn.



42.7a–e

When fashioning the stoma, a limited number of sutures is used to secure the intestine to the anterior abdominal wall to avoid compromising the blood supply. It is not mandatory to secure the serosa to the fascial layer of the abdominal wall, although some surgeons believe that this reduces the risk of stomal prolapse. Similarly, the choice of whether to evert the mucosa as a mature stoma is left to the individual. The mucous fistula is attached to the skin in a similar fashion, but may be left flush with the wound. When the stoma is fashioned away through its own separate "stab wound" type of opening, care must be taken when delivering the bowel through the anterior abdominal wall to ensure that the tract is neither too small (which may lead to stenosis) nor too large (which may predispose to prolapse) and that the bowel passes through *en masse* without stripping of the serosa.

Operations for multifocal necrotizing enterocolitis

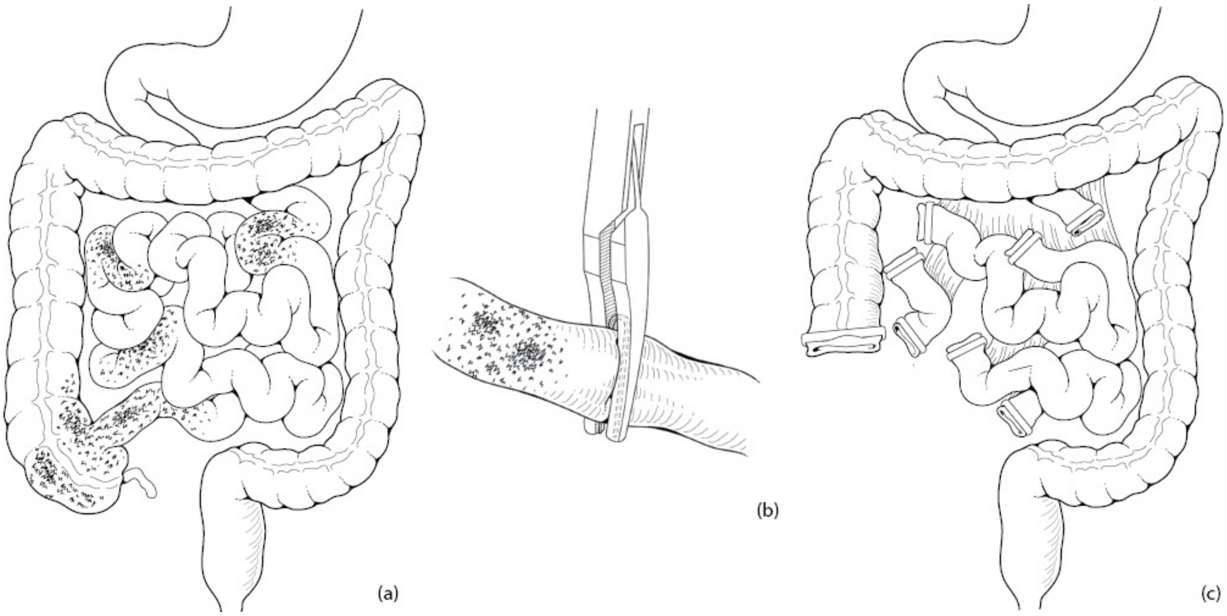
When there are multiple separate segments of diseased intestine separated by lengths of healthy bowel, there are a number of surgical options at laparotomy, depending on the overall condition of the infant, the viability of any potential resection margins, and the mobility of the remaining intestine and mesentery. Of prime concern in such cases is the excision of all gangrenous intestine while preserving as much bowel length as possible. Measuring the remaining healthy intestine may be used as a guide to expected outcome. In a stable child with minimal peritoneal soiling and healthy resection margins, it is possible to perform multiple resections and multiple primary anastomoses. Performing more than two anastomoses appears to be associated with an increased risk of complications and is therefore not advised.

Stoma formation may also be used in such cases of multifocal disease. If this approach is selected, the most proximal resection margin should be brought out as a stoma. In some cases of multifocal disease, a combination of stoma formation and primary anastomosis may be most appropriate. Resection margins suitable for primary anastomosis may be joined and areas of more doubtful viability may be exteriorized as a stoma. Similarly, if an anastomosis has been performed following resection of one segment of diseased intestine but it is not possible to determine the viability of the colon, a stoma may be formed below a primary anastomosis. This approach is preferable to the formation of multiple stomas and requires the closure of the stoma rather than a full laparotomy to restore intestinal continuity at a later stage.

In addition, the “clip and drop” operation may be used in the presence of multifocal disease to preserve as much bowel length as possible.

“CLIP AND DROP” OPERATION

This technique allows for removal of gangrenous intestine and also avoids stoma formation. All segments of grossly non-viable or perforated bowel are resected (Figure 42.8a and b) and the peritoneal cavity irrigated. The ends of remaining bowel are clipped using Ligaclips or tied off circumferentially using a suture and returned to the abdomen (**Figure 42.8c**). This is followed by a second-look laparotomy, ideally with anastomosis, not longer than 72 hours later. This procedure may be combined with stoma formation as appropriate.



42.8a–c

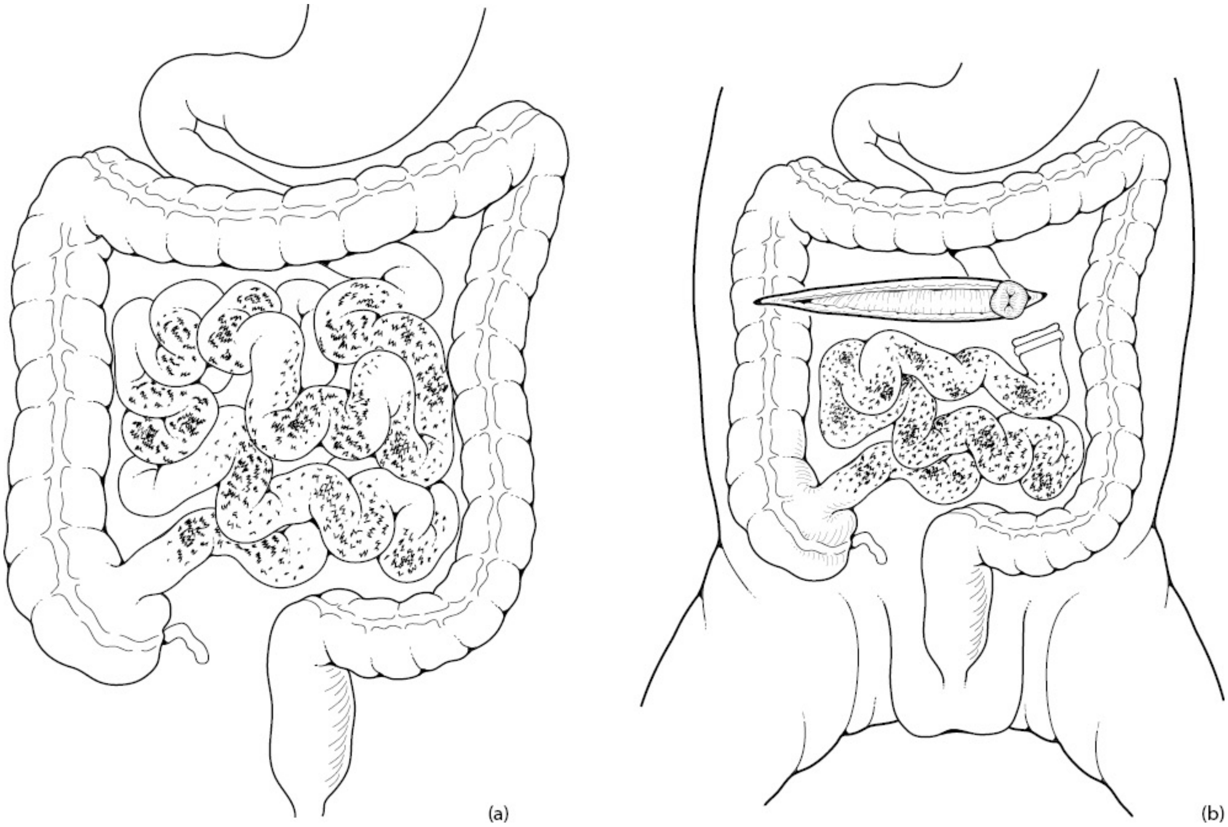
Operations for pan-intestinal necrotizing enterocolitis

The techniques described thus far are of particular use for the infant with one or more short segments of NEC. Infants with NEC affecting a large proportion of the GI tract pose a particularly difficult problem, and treatment of this group remains controversial. The surgical principles in these children are difficult if not impossible to fulfill. Due to the length of bowel involved, it is often not possible fully to remove all gangrenous intestine while salvaging adequate length for sustainable life. It is for this reason that in the infant with pan-intestinal NEC who is unstable and critically ill, some surgeons would perform an “open and close” laparotomy, ascertaining the futility of further treatment with subsequent withdrawal of therapy. However, when there is doubt, a number of techniques may be utilized with the aim of allowing time for stabilization of the infant’s general condition and the chance of some healing of the GI tract to occur. Due to the severity of the disease, the mortality in these cases remains high regardless of the surgical approach. The procedures available are the “clip and drop” (as described earlier) and “patch, drain, and wait” approaches, and the formation of a proximal defunctioning jejunostomy.

PROXIMAL JEJUNOSTOMY

Surgical creation of a proximal jejunostomy in the presence of pan-intestinal disease allows decompression and defunctioning of the diseased intestine but does not remove gangrenous segments and may permit continued bacterial translocation. This procedure is useful in neonates with NEC affecting the majority of the intestine, but the high morbidity and mortality rates should be carefully considered.

The jejunum is divided proximally to the most diseased section of intestine and the distal resection margin oversewn. The proximal margin is exteriorized as a stoma as previously described, either through the end of the wound or at a separate site (Figure 42.9a and b).



42.9a,b

Once the infant is more stable, the intestine has had a chance to recover from the initial insult, and the infant has had the opportunity to grow, a second laparotomy is performed. A definitive procedure is undertaken and the stoma closed. A specific complication of forming a high jejunostomy is the risk of massive stomal output resulting in fluid and electrolyte imbalance. This may expedite the need for second laparotomy and stomal closure.

POSTOPERATIVE CARE

The postoperative care of infants with NEC is invariably on the neonatal intensive care unit with continuous monitoring. Respiratory support is usually necessary and cardiovascular support often is. A systemic inflammatory response often develops in response to either NEC or surgery (or a combination of both), and failure of one or more organ systems may ensue, requiring appropriate intervention. Sepsis is a common problem during the postoperative period, despite the routine use of broad-spectrum antibiotics for 10 days. Feeds are reintroduced 10 days following surgery, using a cautious regime and increasing as tolerated.

COMPLICATIONS

Of principal concern in the immediate postoperative period is the infant's survival, typically not as a result of the surgery itself but related to the severity of the underlying disease and degree of organ dysfunction. Beyond this period, infants who have survived an episode of NEC continue to have a higher than anticipated mortality both prior to discharge from the neonatal intensive care unit and within the first year of life, primarily due to coexisting morbidity related to prematurity.

For reasons that are not immediately apparent, as many as 10% of infants may suffer a second episode of NEC and a small number are troubled by further recurrent disease. Such infants are at high risk of developing intestinal failure, either due to a need for extensive intestinal resection or as a consequence of functional intestinal failure, the pathophysiology of which is poorly understood.

Infants who do not tolerate feeds or who initially tolerate feeds but subsequently develop an intolerance often with abdominal distension and/or frequent loose stools and failure to thrive should be investigated for the presence of an intestinal stricture. These may occur at a site of non-resected NEC or at the site of an intestinal anastomosis. In suspected cases, a GI contrast study may prove the diagnosis. Treatment is by laparotomy, with resection of the strictured intestine and primary anastomosis.

Closure of stoma

Once the infant is thriving and has achieved satisfactory weight gain, any stoma may be closed and intestinal continuity restored. Typically, this is after a minimum period of 6 weeks but some surgeons prefer to wait longer for further infant growth and development before stoma closure. Earlier stoma closure may be of benefit in preterm infants who frequently have difficulty in thriving with a stoma. Prior to closing any stoma, a contrast study should be undertaken of the distal intestine to ensure the absence of intestinal stricture requiring resection. The stoma is closed by performing an anastomosis between the two ends of bowel in an identical fashion to that described for primary anastomosis in the setting of acute NEC.

OUTCOME

NEC appears to be associated with a significant long-term morbidity, although the long-term outcome for infants with NEC is poorly reported. The most serious GI complication of NEC is short bowel syndrome, with an incidence of up to 23% in NEC survivors. Neurodevelopmental implications of NEC are being increasingly recognized as important although the mechanisms responsible remain to be elucidated. Only approximately 50% of the neonates with NEC are neurodevelopmentally normal. Further research is required to identify causative factors and improve neurodevelopmental outcome in this group of infants.

The mortality rate of neonates with NEC depends on the severity of the disease, associated anomalies, weight, and gestational age. Overall mortality in a recent epidemiological study of study of 189 neonates requiring laparotomy for NEC was 15% within 28 days of initial procedure, rising to 22% by 1 year of age. In survivors, a high proportion required further surgery within the first year of life. There is little doubt that these figures are significantly worse in extremely low birth weight infants born at <1000 g. Overall, while the surgical outcome of treatment for NEC may be successful in the majority of cases, the long-term morbidity and mortality associated with such premature infants should not be overlooked.

FURTHER READING

Allin B, Long A-M, Gupta A, et al. BAPS-CASS Necrotising Enterocolitis Collaboration – A UK-wide cohort study describing management and outcomes for infants with surgical necrotising enterocolitis. *Sci Rep* 2017; 7: 41149.

Fitzgibbons CS, Ching Y, Yu D, et al. Mortality of necrotizing enterocolitis expressed by birth weight categories. *J Pediatr Surg* 2009; 44: 1072–6.

Fullerton BS, Hong CR, Velazco CS, et al. Severe neurodevelopmental disability and healthcare needs among survivors of medical and surgical necrotizing enterocolitis: A prospective cohort study. *J Pediatr Surg* 2017; S0022-3468(17)30651-6.

Haricharan RN, Gallimore JP, Nasr A. Primary anastomosis or ostomy in necrotizing enterocolitis? *Pediatr Surg Int* 2017; 33: 1139–45.

Moss RL, Dimmitt RA, Barnhart DC, et al. Laparotomy versus peritoneal drainage for necrotizing enterocolitis and perforation. *N Engl J Med* 2006; 354: 2225–34.

Rees CM, Eaton S, Kiely EM, et al. Peritoneal drainage or laparotomy for neonatal bowel perforation? A randomized controlled trial. *Ann Surg* 2008; 248: 44–51.

Yikilmaz A, Hall NJ, Daneman A, et al. Prospective evaluation of the impact of sonography on the management and surgical intervention of neonates with necrotizing enterocolitis. *Pediatr Surg Int* 2014; 30: 1231–40.

43

Anorectal malformation

MARC A. LEVITT AND RICHARD J. WOOD

HISTORY

Anorectal malformations (ARMs) have been described for centuries, with most children undergoing an operation to create an anal orifice on the perineum. If the rectum was located very close to the skin, most children survived, but many died, probably because the rectum was unreachable, located high in the pelvis. In 1835, Amussat reported, for the first time, suturing of the rectal wall to the skin edges, essentially the first anoplasty. Thereafter, surgeons performed a perineal operation, without a colostomy, for “low malformations.” “High imperforate anus,” on the other hand, was usually treated in the newborn period with a colostomy, followed by an abdominoperineal pull-through (with rectal resection) later in life. The specific recommendation was to pull the intestine as close to the sacrum as possible to avoid trauma to the genitourinary tract. Douglas Stephens performed the first objective anatomic studies of human cadavers with these defects, and in 1953 proposed an initial sacral approach to separate the rectum from the urinary tract with preservation of the puborectalis sling (considered a key factor in maintaining fecal continence). He also suggested opening the abdomen, if necessary, after the sacral approach.

Following Stephens’ recommendations, several different surgical techniques were proposed. The common denominator in all of these techniques, most notably the Kieswelter technique, was the protection and utilization of the puborectalis sling. Justin Kelly brought the Stephens approach to Boston Children's Hospital in the late 1960s where Alberto Peña and he were trainees. Then, from 1972 to 1980, once back in Mexico, Peña made the perineal incision longer and longer to improve the exposure, and in collaboration with Peter Devries performed a posterior sagittal anorectoplasty for the first time. This approach allowed direct exposure of this difficult-to-reach anatomic area by incising to find the rectum and then reconstructing the funnel-like sphincter mechanism around the mobilized rectum. It then became possible to correlate the external appearance of the perineum with the operative findings and the clinical results. The approach had dramatic implications for understanding the anatomy of these defects, terminology, classification, and most importantly, treatment, and essentially launched the field of pediatric colorectal surgery. Thereafter, major advances including the use of laparoscopy introduced by Keith Georgeson and application of the posterior sagittal approach to other situations (such as cloaca) emerged.

PRINCIPLES AND JUSTIFICATION

Incidence

ARMs occur in 1 in 4000 of neonates, slightly more commonly in boys than in girls. The most common defects in girls are a rectovestibular fistula followed by a rectoperineal fistula. A true rectovaginal fistula is rare. Most of the "rectovaginal fistulas" described are probably cases of misdiagnosed cloacas or rectovestibular fistulas. The third most common defect in girls is a cloaca.

The most common defect in boys is a rectourethral fistula, followed by a rectoperineal fistula. Recto-bladder neck fistulas in boys represent 10% of the entire group of defects. Imperforate anus without fistula in both boys and girls occurs in only 5% of the entire group of defects, although it is particularly common in patients who have Down syndrome. The estimated risk of having a second child with an ARM is 1.4%, but, when the first child is born with a perineal or vestibular fistula, this incidence increases to 3%. There is less family transmission among patients with cloacas, recto-bladder neck, and rectourethral prostatic fistulas.

Classification

The anatomic classification of ARMs is shown in Table 43.1.

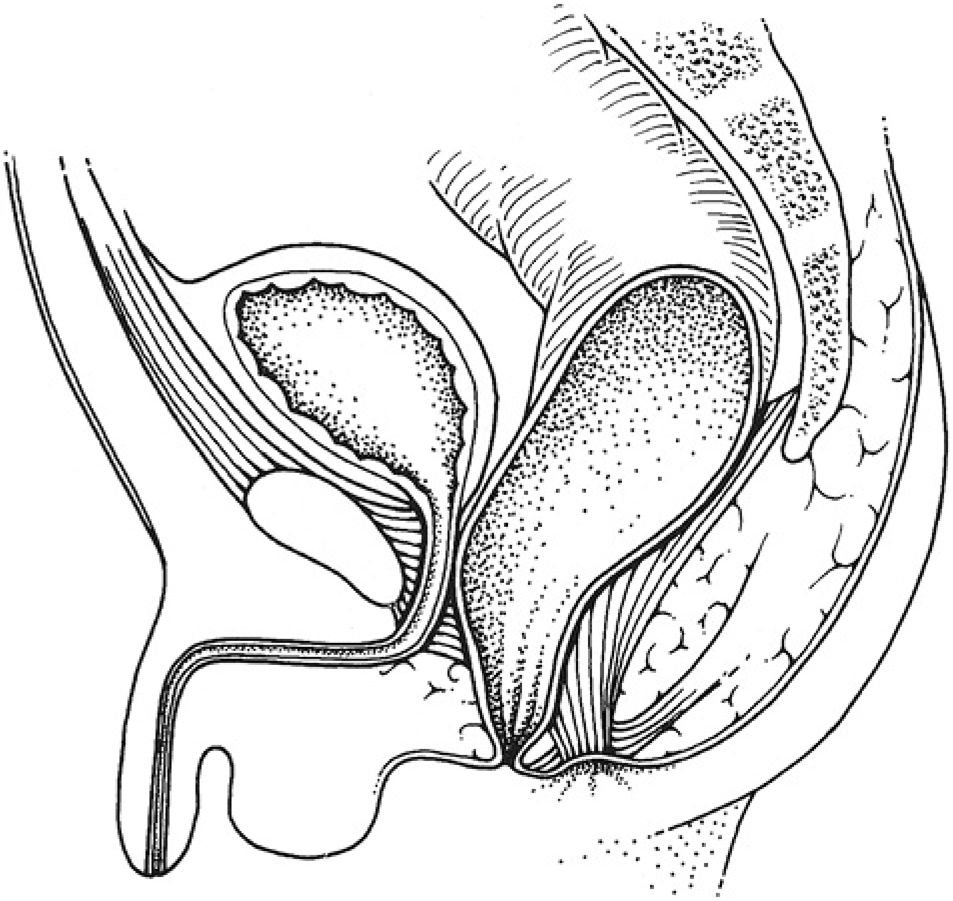
Table 43.1 **Classification of anorectal malformations**

Males	Females
Anal stenosis	Anal stenosis
Rectal atresia	Rectal atresia
Rectoperineal fistula	Rectoperineal fistula
Imperforate anus without fistula	Imperforate anus without fistula
Rectourethral fistula	Vestibular fistula
• Bulbar	Cloaca
• Prostatic	
• Recto-bladder neck fistula	

MALES

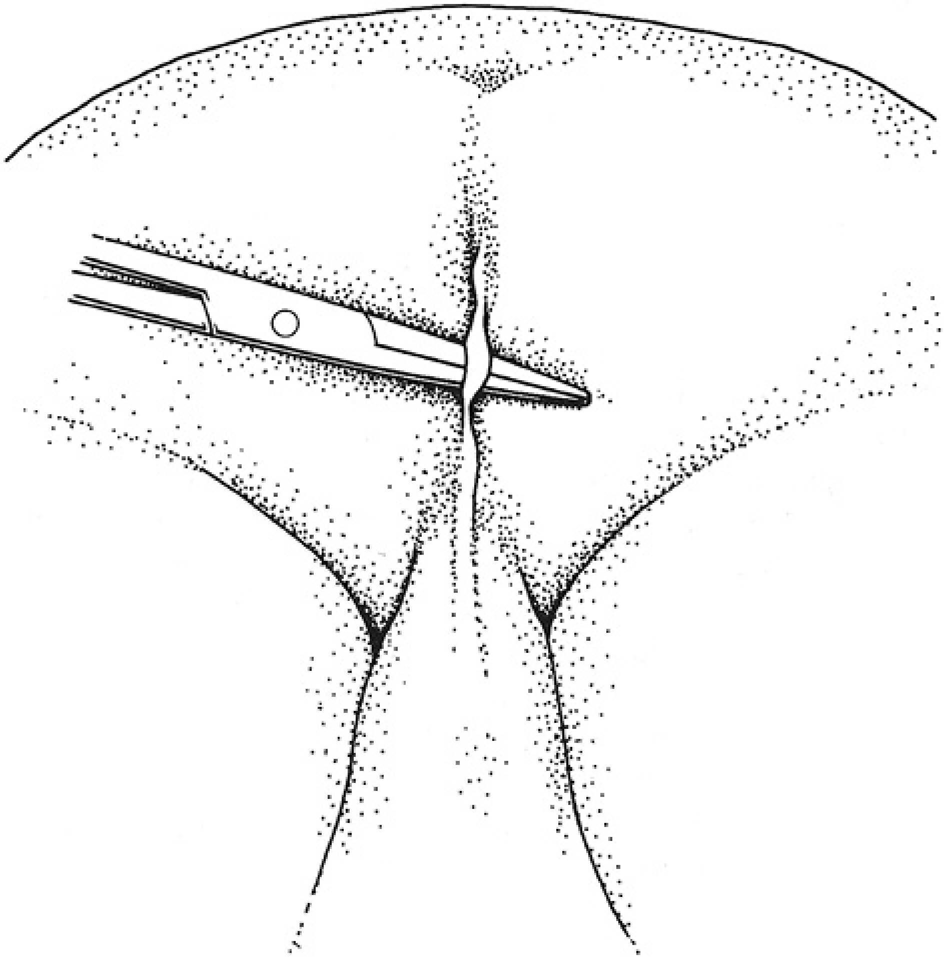
Rectoperineal fistula

In this defect, the rectum is located within most of the sphincter mechanism. Only the lowest part of the rectum, which is fistulous tissue, is anteriorly displaced and lies outside the sphincter mechanism (Figure 43.1).



43.1

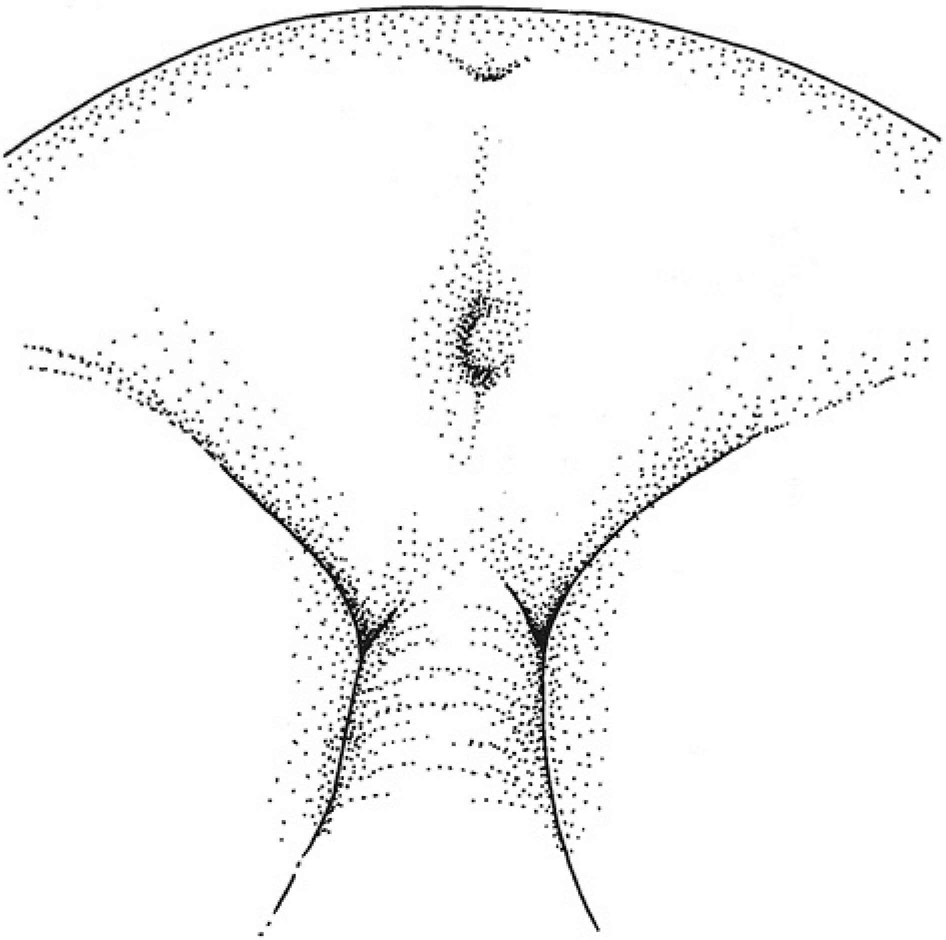
Sometimes, the fistula follows a subepithelial midline tract opening along the midline perineal raphe, scrotum, or penis. The perineal findings in this kind of defect include a prominent skin tag, below which an instrument can be passed, known as a “bucket-handle” malformation (**Figure 43.2**), a black (meconium) or white (mucous), ribbon-like, midline structure that represents a subepithelial fistula or a very well-formed anal dimple suggesting the presence of a very low defect. The diagnosis is established by perineal inspection.



43.2

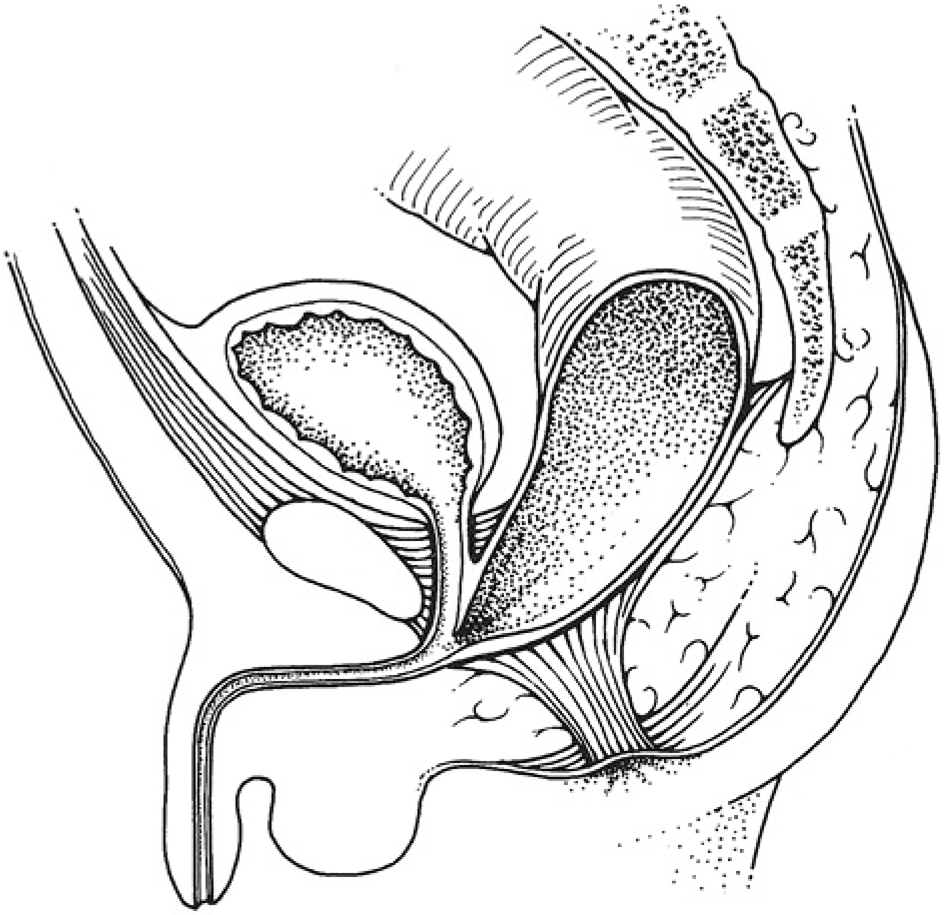
Rectourethral fistula

This is the most common defect in boys and appears with an anal dimple and no obvious fistula (**Figure 43.3**)

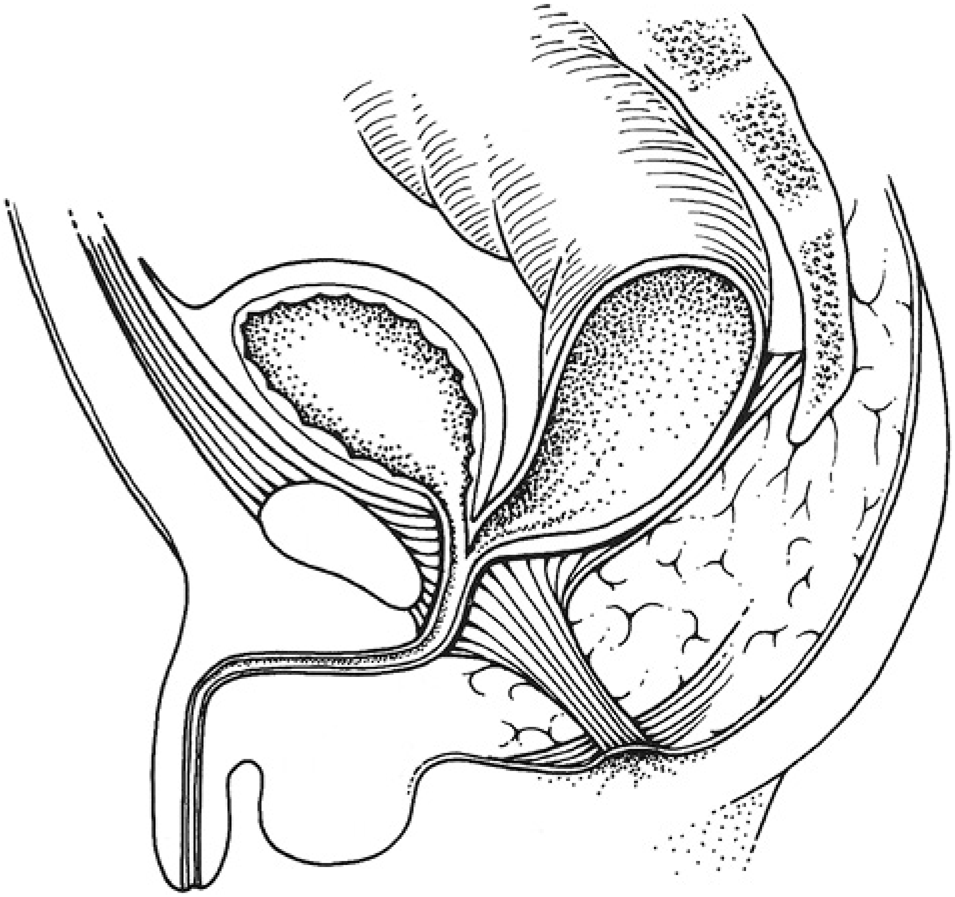


43.3

The rectum may communicate with the lower part of the urethra (bulbar urethra, **Figure 43.4**) or with the upper urethra (prostatic urethra (**Figure 43.5**). Immediately above the fistula site, the rectum and urethra share a common wall with no plane of dissection. The lower the rectum, the longer is this common wall. The rectum is surrounded laterally and posteriorly by the levator muscles, and between the end of the rectum and the perineal skin there is a portion of voluntary muscle called the "muscle complex." The contraction of the levator muscle pushes the rectum forward. The contraction of the muscle complex elevates the skin of the anal dimple. At the level of the skin, and located on both sides of the midline, there is a group of voluntary muscle fibers called "parasagittal fibers."

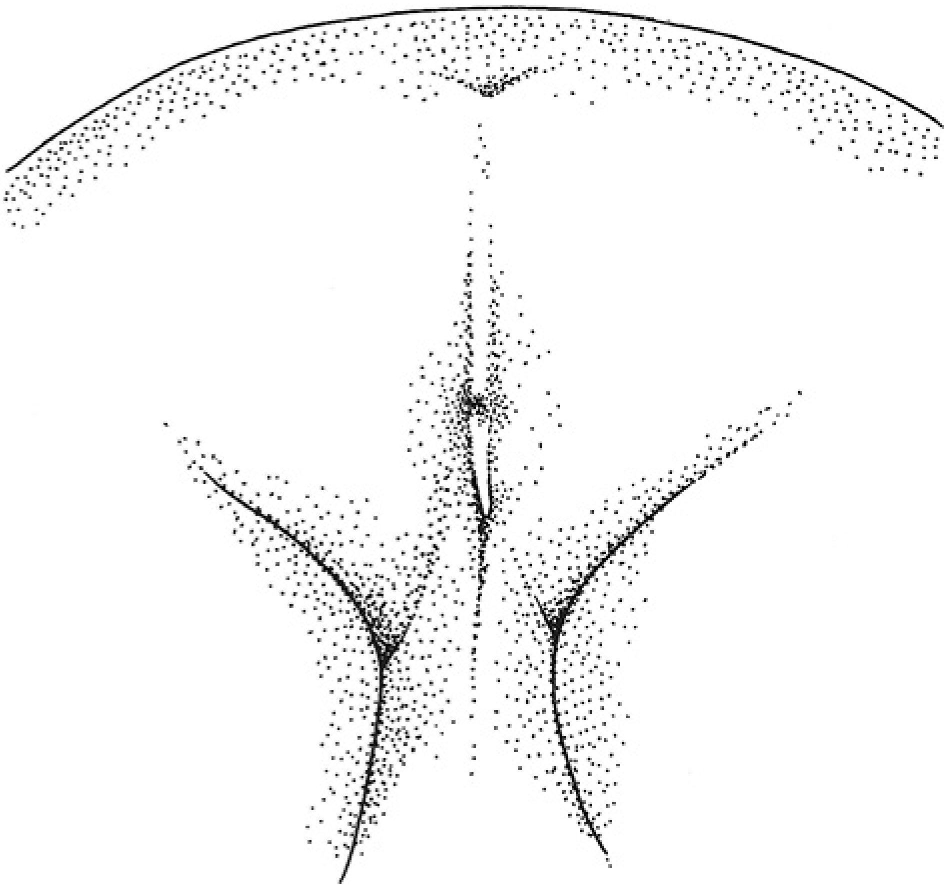


43.4



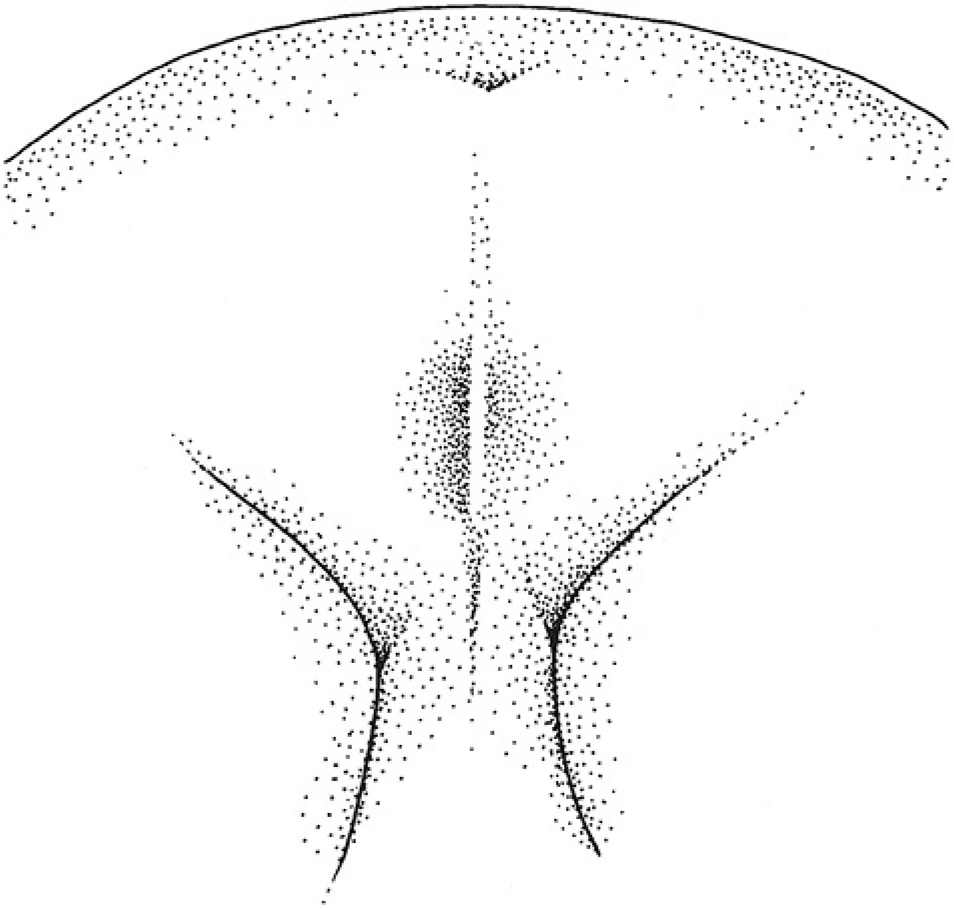
43.5

Patients with rectourethral bulbar fistulas usually have a normal sacrum and a perineum with a prominent midline groove (**Figure 43.6**).



43.6

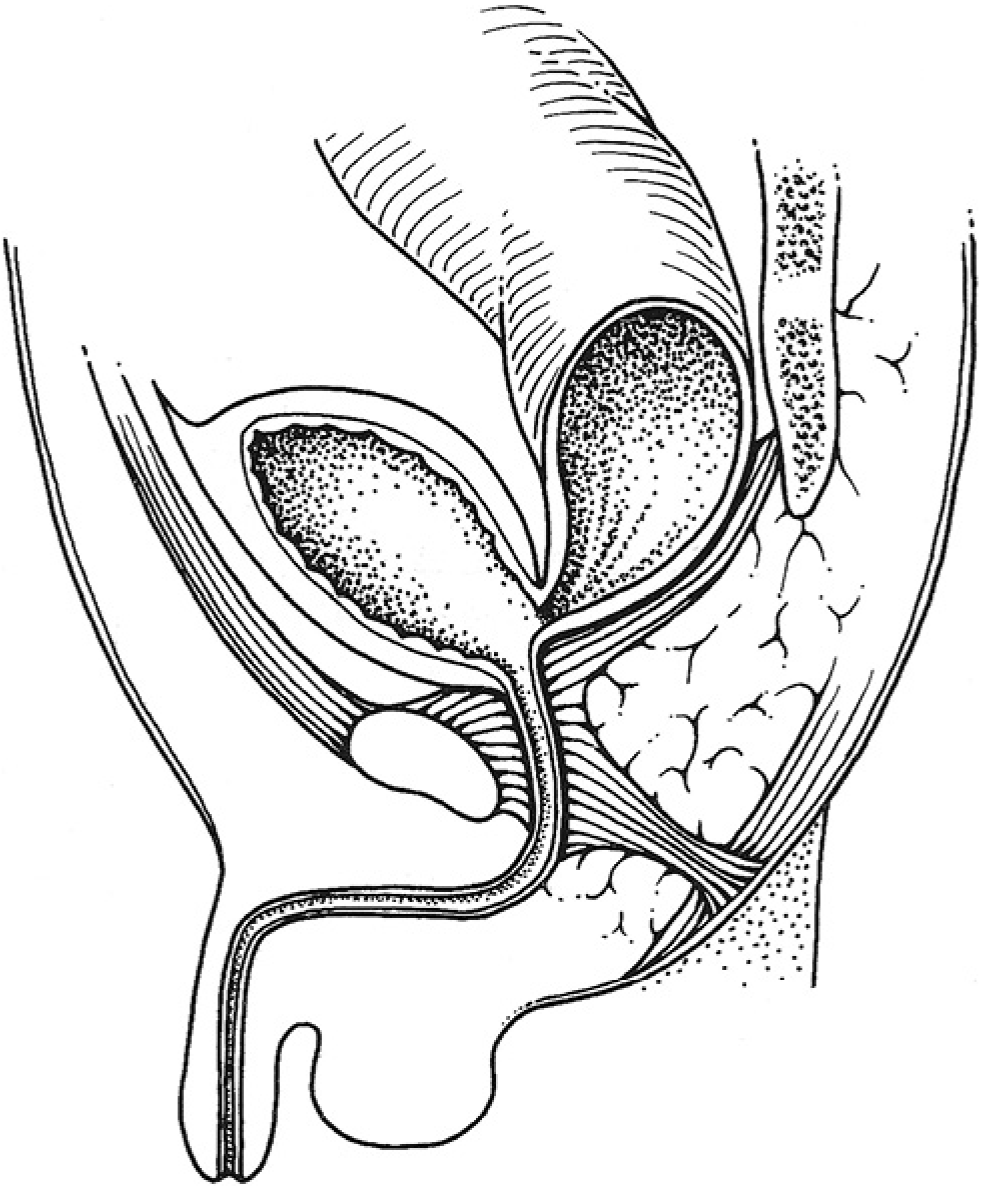
Patients with a rectourethral prostatic fistula sometimes have an abnormal sacrum, underdeveloped sphincter mechanism, and flat perineum (**Figure 43.7**). The anal dimple is often located very close to the scrotum. Neonates with rectourethral fistulas may pass meconium through the urethra, usually after 20 hours of life, which is an unequivocal sign of a rectourethral fistula.



43.7

Recto-bladder neck fistula

In these malformations, the rectum communicates with the urinary tract at the bladder neck (**Figure 43.8**). The levator muscle, muscle complex, and parasagittal fibers are often poorly developed and the sacrum deformed or absent. The entire pelvis seems to be underdeveloped (caudal regression), and its anteroposterior diameter is foreshortened. The perineum is usually flat. For all these reasons, the prognosis for bowel control is usually poor.



43.8

Imperforate anus without fistula

In these cases, the rectum is blind and is almost always found at the same level as in cases with rectourethral bulbar fistula. The sacrum and sphincteric mechanism are usually normal and therefore these patients have a good prognosis. This is a common malformation seen in patients with Down syndrome.

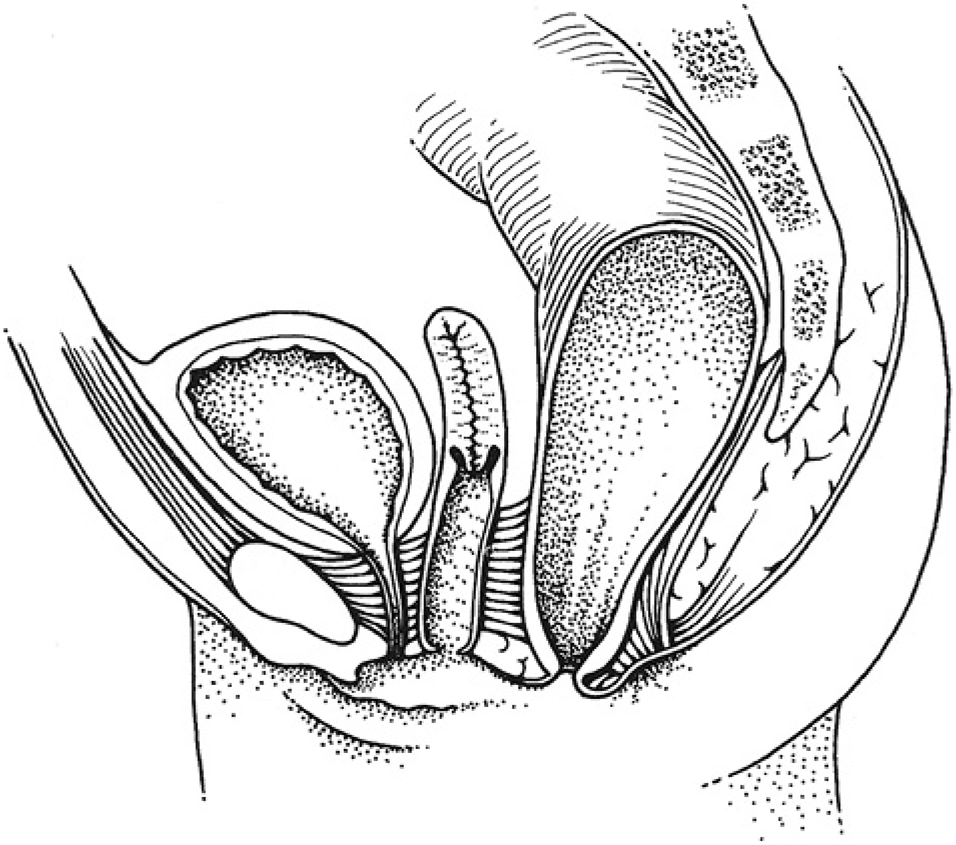
Rectal atresia/stenosis

This is a very unusual defect, occurring in only 1% of cases. These are the only patients with imperforate anus who have a normal appearing anal canal. Externally, the anus looks normal, with a somewhat funnel-like appearance, and the malformation is often discovered after the onset of low intestinal obstruction or related to severe constipation. About 2 cm from the anal verge, there is an atretic or stenotic area. The upper blind rectum is usually located very close to the anal canal. The sacrum is normal, but can be a hemisacrum. The sphincteric mechanism is excellent, and therefore the prognosis is good. This malformation is associated with a presacral mass, usually a teratoma or meningocele.

FEMALES

Rectoperineal fistula

This defect is similar to the rectoperineal fistula described for boys. The rectum and vagina are separated (**Figure 43.9**). The sphincteric mechanism and therefore the functional prognosis for bowel control are also good.

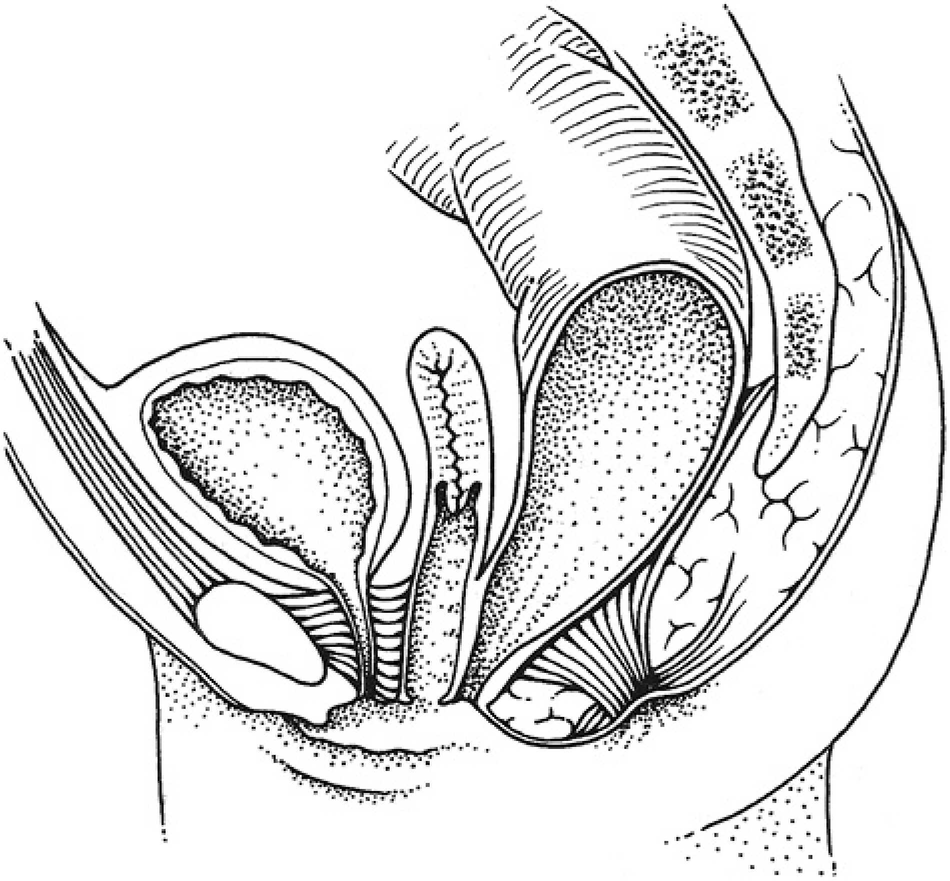


43.9

Rectovestibular fistula

This is the most common defect seen in girls. It has an excellent functional prognosis, but unfortunately is the most common type to suffer a failed repair.

The distal rectum opens in the vestibule of the female genitalia immediately posterior to the hymen. Immediately above the fistula site, the rectum and vagina share a thin common wall (**Figure 43.10**). The diagnosis is established by perineal inspection. These patients usually have good muscles and a normal sacrum. Sometimes they are mislabeled as having a rectovaginal fistula, an imprecise description of the newborn genitalia.



43.10

Imperforate anus without fistula and rectal atresia/stenosis

These defects in girls have the same anatomic characteristics as those described in boys, and therefore have similar prognostic implications.

ASSOCIATED DEFECTS

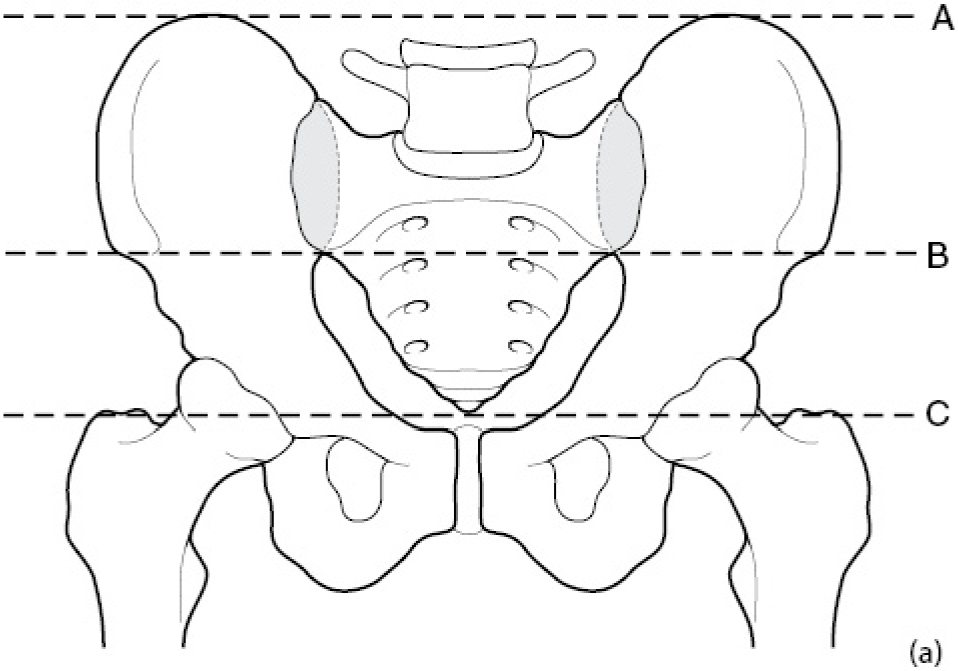
Sacrum and spine

The sacrum is often abnormal in ARMs. There appears to be good correlation between the degree of sacral development and the final functional prognosis.

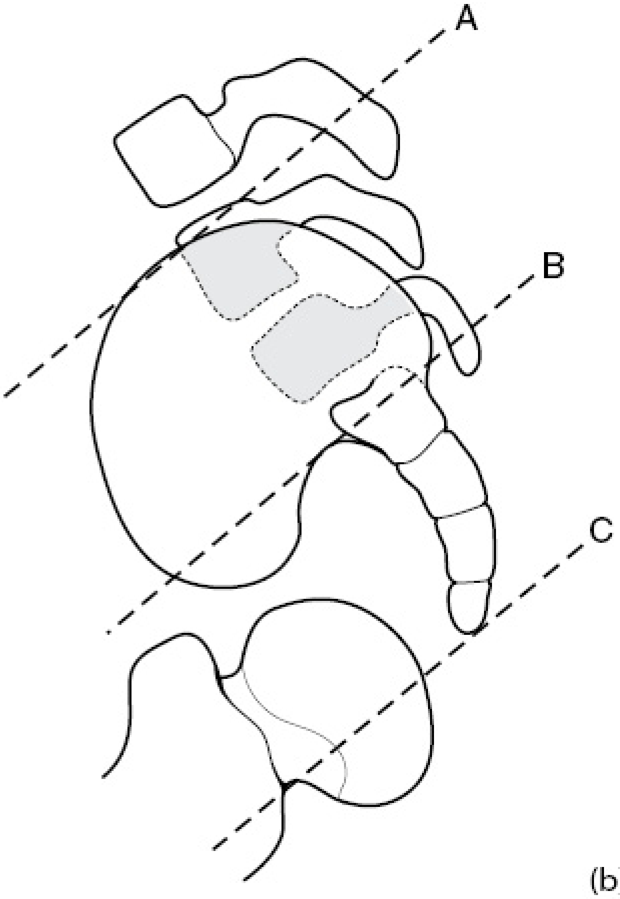
A sacral ratio can express the degree of sacral development. For this measurement, three lines are drawn (**Figure 43.11a and b**):

- Line A extends across the uppermost portion of the iliac crests.

- Line B joins both inferior and posterior iliac spines and passes through the sacroiliac joint.
- Line C runs parallel to lines A and B and passes through the lowest radiologically visible sacral point.



(a)

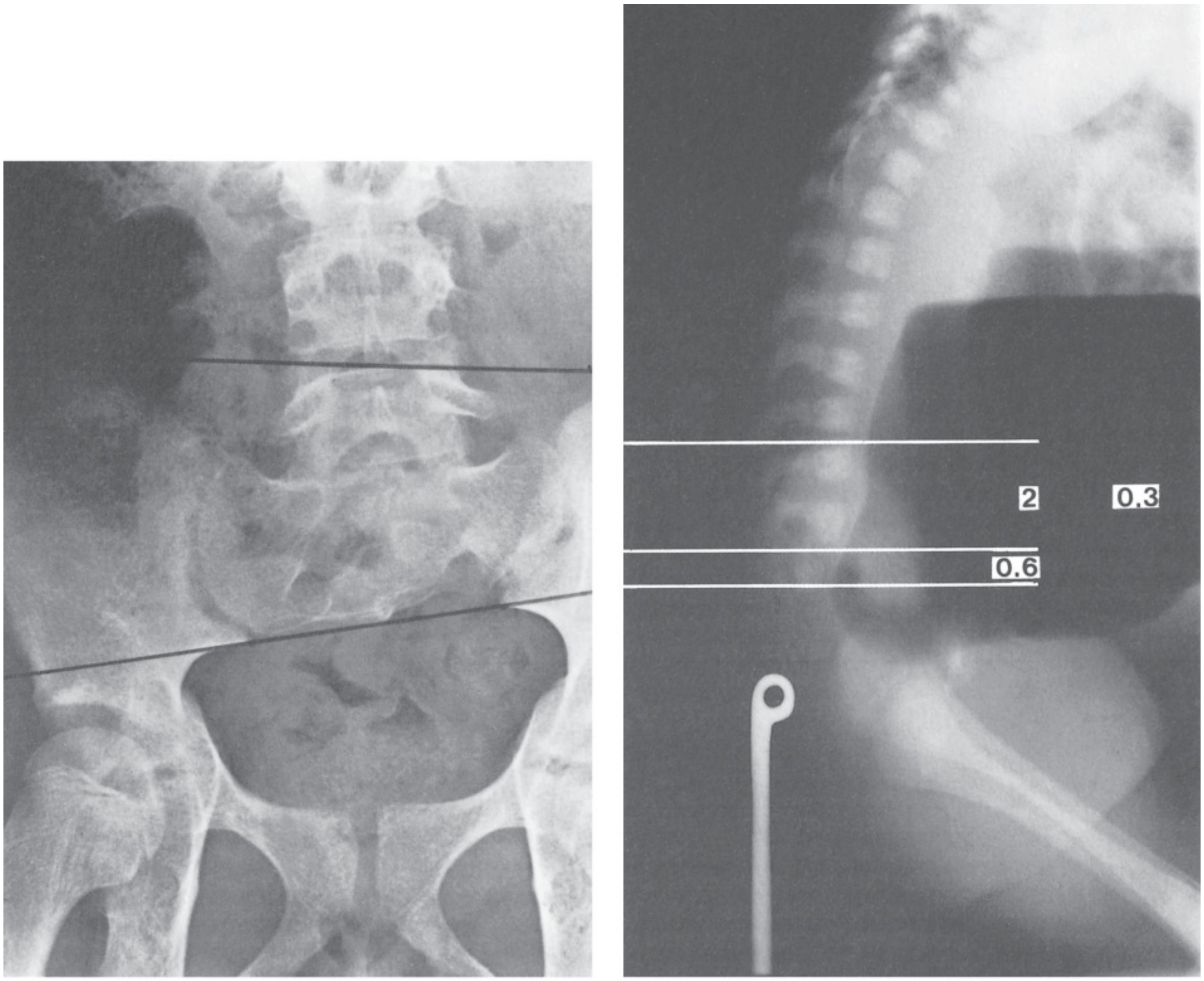


(b)

43.11a,b

In normal children, the ratio of the distances BC : AB lies between 0.7 and 0.8 in both anteroposterior and lateral projections. Children with ARMs have a degree of sacral hypodevelopment, with the ratio varying between 0 and 1.0. A ratio of less than 0.4, for example, represents a poor functional prognosis.

Two different examples of patients with poor sacral ratios are shown in **Figure 43.12a and b**.



(a) (b)

43.12a,b

Vertebral anomalies may include hemivertebrae located in the lumbar or thoracic spine. The prognostic implications of these types of defect in terms of bowel and urinary control are not known. These patients often need to be followed for scoliosis. All neonates with anorectal anomalies require investigation of their spinal canal, generally with a spinal ultrasound, to assess for tethered cord, with potentially follow-up magnetic resonance imaging (MRI) at 6–12 months. With the knowledge of the type of ARM, the sacral ratio, and the status of the spine, the clinician can make a good prediction of bowel control. These three factors when assessed together comprise the ARM continence index (**Table 43.2**).

Table 43.2 **ARM Continence Index components**

1. ARM type
    - Rectoperineal fistula
    - Rectal stenosis
    - Rectal atresia
    - Recto bulbar fistula
    - Recto vestibular fistula
    - Imperforate anus without fistula
    - Cloaca <3 cm common channel
    - Recto prostatic fistula
    - Recto vaginal fistula
    - Recto-bladder neck fistula
    - Cloaca >3 cm common channel
    - Covered exstrophy
    - Cloacal exstrophy
2. Spine
    - Termination (end) of the conus
        - Normal
        - Abnormally low termination
    - Myelomeningocele
        - Yes
        - No
    - Filum appearance
        - Normal
        - Abnormal – fatty thickening
        - Myelomeningocele
3. Sacrum
    - Sacral ratio
        - Greater than 0.7
        - Between 0.4 and 0.69
        - Less than 0.4
    - Hemisacrum
    - Presacral mass
    - Sacral hemivertebra

Urologic and gynecologic defects

Associated urogenital defects are common and the reported variation may reflect the accuracy and thoroughness of the urologic and gynecologic investigations. Patients with cloaca or recto-bladder neck fistula have a 90% chance of having significant associated urologic abnormality. Children with more minor defects (rectoperineal fistula) have less than a 10% chance of having an associated urologic defect. The most common urologic malformation is absent kidney, followed by vesicoureteric reflux. Hydronephrosis, urosepsis, and metabolic acidosis from poor renal function represent the main sources of morbidity in children with ARMs. All need an ultrasonographic study of the abdomen done during the first 24 hours after birth. If this study shows some abnormalities, a thorough urologic evaluation is indicated.

Gynecologic issues, such as a vaginal septum and absent vagina, are common (5–10% of rectovestibular fistulas), and inspection of the vaginal canal is important prior to proceeding with surgical intervention. In cloacas, duplicated Müllerian systems and hydrocolpos occur in 30–40% of cases.

Other defects

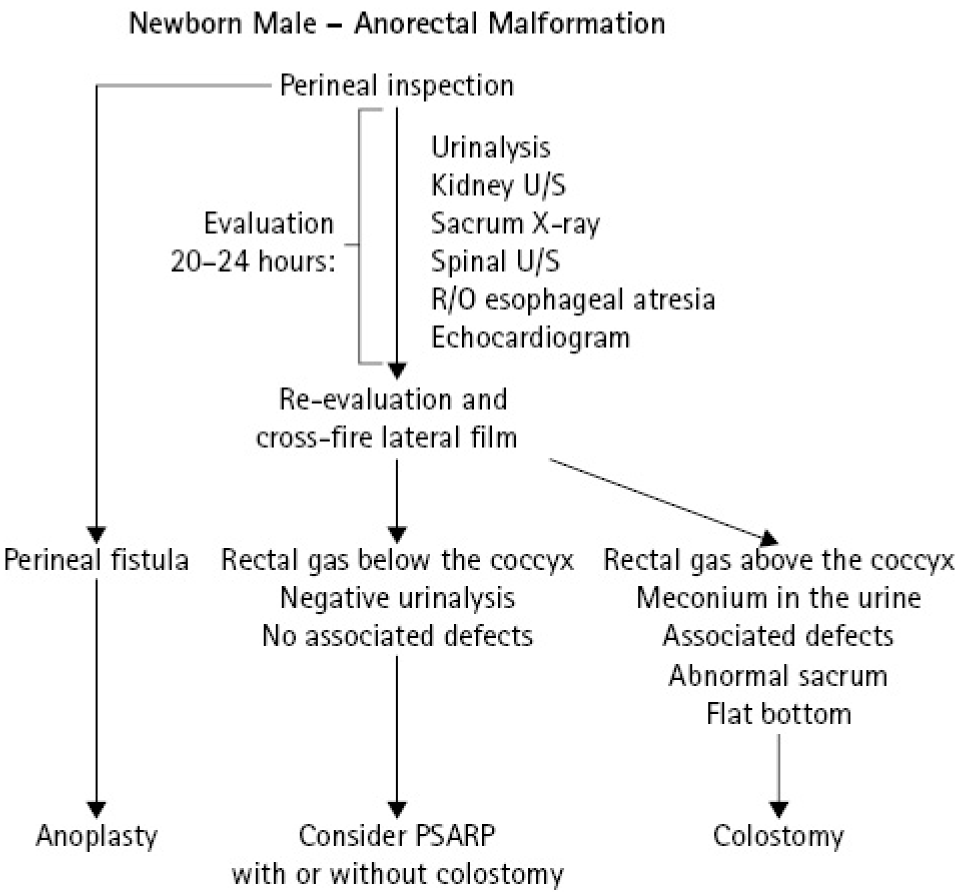
Other congenital malformations are commonly associated with ARMs including esophageal atresia, duodenal atresia, malrotation, and cardiovascular defects.

MANAGEMENT OF ARMS DURING THE NEONATAL PERIOD

Two important questions must be answered during the first 24 hours of life: What are the associated anomalies, and what operation is required, a newborn pull-through or a colostomy?

Males

The decision-making algorithm for the management of newborn males with ARMs is shown in **Figure 43.13**. Associated malformations should be looked for. In most boys, perineal inspection and checking the urine for meconium will provide enough clinical evidence to make an anatomic diagnosis. If a rectoperineal fistula exists, the patient may be treated with an anoplasty in the newborn period. The presence of a flat bottom and meconium in the urine are indications for a colostomy, which decompresses the intestine in the neonatal period, provides access for a contrast study to define the anorectal anatomy, and will subsequently provide protection against infection during the healing process after the main anorectal repair.


Newborn Male – Anorectal Malformation
Perineal inspection
Evaluation 20–24 hours:
Urinalysis
Kidney U/S
Sacrum X-ray
Spinal U/S
R/O esophageal atresia
Echocardiogram
Re-evaluation and cross-fire lateral film
Perineal fistula
Rectal gas below the coccyx
Negative urinalysis
No associated defects
Rectal gas above the coccyx
Meconium in the urine
Associated defects
Abnormal sacrum
Flat bottom
Anoplasty
Consider PSARP with or without colostomy
Colostomy

43.13 Decision-making algorithm for the management of newborn males with anorectal malformation.

It is important to wait 20–24 hours before making any decision about surgical intervention because these patients do not show abdominal distension during the first few hours of life. Even if a rectoperineal fistula is present, meconium is not usually seen on the perineum right after birth because a significant amount of intra-abdominal pressure is required for the meconium to be forced through a perineal or urinary fistula and to overcome the voluntary muscle tone that keeps the most distal part of the rectum compressed (**Figures 43.1, 43.4, and 43.5**). Radiologic evaluations performed during the first hours of life are therefore unreliable, as they will show a falsely high rectum.

Shortly after birth, IV fluids should be administered and a nasogastric tube inserted to keep the stomach decompressed, avoid the risk of vomiting and aspiration, and assess for an associated esophageal atresia. An ultrasound of the abdomen is performed to check for other anomalies (mainly urologic). A piece of gauze is placed over the penis to check for particles of meconium.

If after a day of observation there is no clinical evidence indicating the need for a colostomy or demonstrating that a primary perineal operation can be safely performed, the patient should have a cross-table lateral film (cross-fire) performed in the prone position to determine the location of the distal rectum. The anal dimple is marked with radio-opaque material. If the rectum is visible below the coccyx, the patient can undergo a primary newborn repair, provided the surgeon is experienced with this technique. If the image is questionable, it is preferable to perform a colostomy.

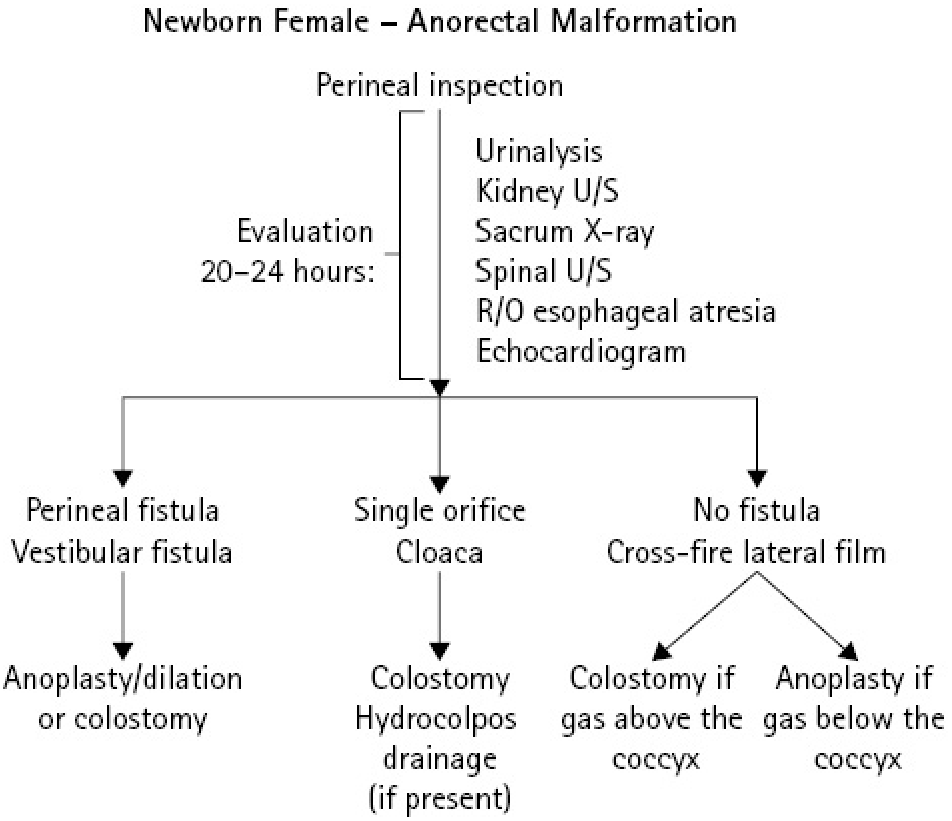
After recovering from the colostomy, the patient is discharged from the hospital. If the infant is growing well and has no other associated defects (cardiovascular or gastrointestinal) that require treatment, they can undergo a distal colostogram to define the distal rectal anatomy at 1–3 months of age, and then a posterior sagittal anorectoplasty.

Performing the definitive repair at that young age has some advantages, including less time with an abdominal stoma, less size discrepancy between proximal and distal stoma at the time of colostomy closure, and anal dilatations, if these are needed that are easier to do. In addition, at least theoretically, placing the rectum in the right location early in life may be an advantage in terms of acquired local sensation.

Some surgeons have proposed a primary repair of all ARMs during the neonatal period without a colostomy. This approach has the potential of avoiding the morbidity related to the formation and closure of a colostomy. However, diagnostic tests (other than a distal colostogram, which obviously requires the presence of a colostomy) used to determine the level of the defect are not accurate enough, and the surgeon could be subjecting the patient to a blind exploration of the perineum. If the rectum is located high in the abdomen, the surgeon may damage other structures including the posterior urethra, seminal vesicles, vas deferens, and ectopic ureters, during the search for the rectum. In addition, there is a risk of dehiscence and infection because the stool is not diverted.

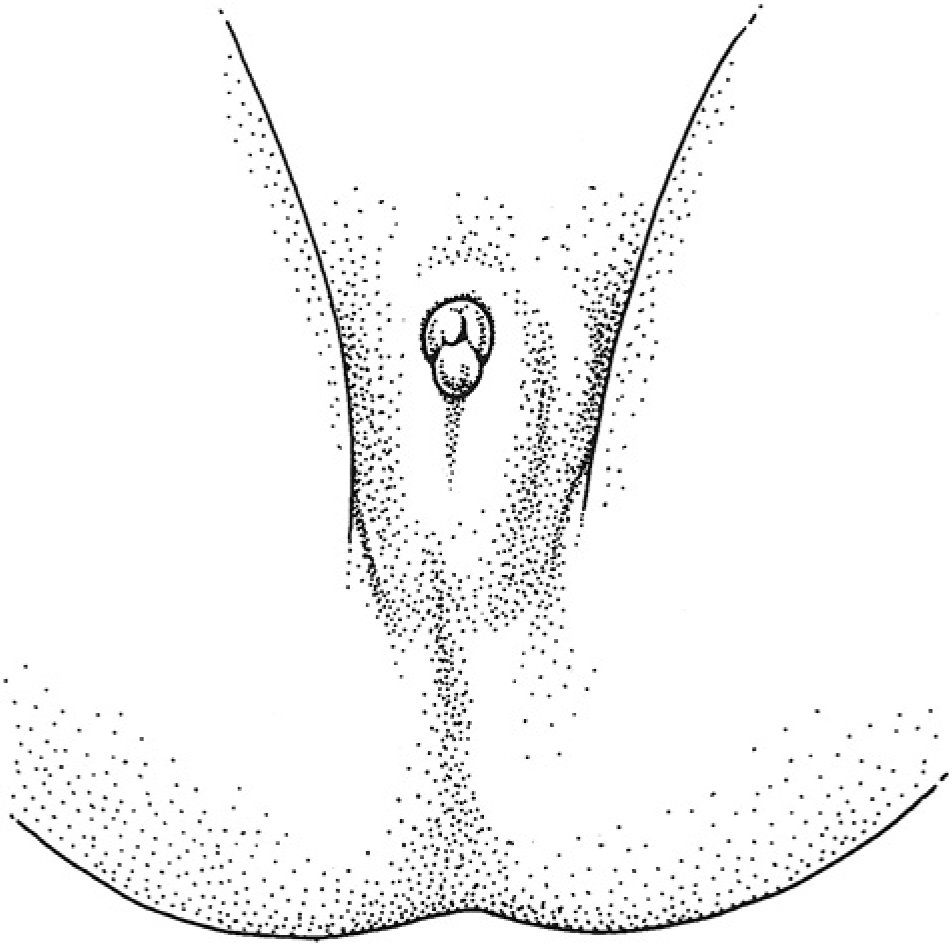
Females

A decision-making algorithm for the initial management of female newborns is shown in **Figure 43.14**. Perineal inspection usually provides more information in girls than in boys. The principle of waiting 20–24 hours before making a decision is again valuable.


Newborn Female – Anorectal Malformation
Perineal inspection
Evaluation 20–24 hours:
Urinalysis
Kidney U/S
Sacrum X-ray
Spinal U/S
R/O esophageal atresia
Echocardiogram
Perineal fistula
Vestibular fistula
Single orifice
Cloaca
No fistula
Cross-fire lateral film
Anoplasty/dilation or colostomy
Colostomy
Hydrocolpos drainage (if present)
Colostomy if gas above the coccyx
Anoplasty if gas below the coccyx

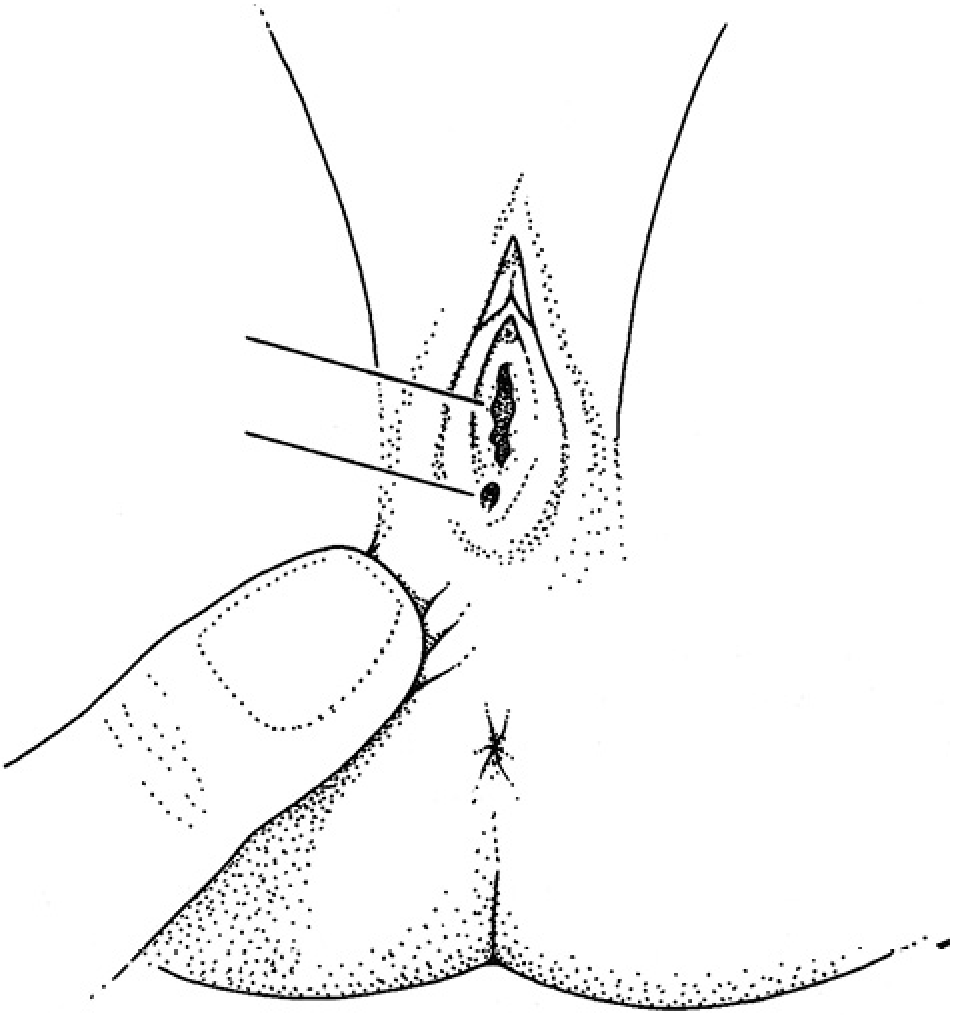
43.14 Decision-making algorithm for the management of newborn females with anorectal malformation.

The presence of a single perineal orifice is pathognomonic for a cloaca (**Figure 43.15**). Because of their complexity, these defects are dealt with separately in Chapter 44.



43.15

Perineal inspection may reveal a rectovestibular fistula, which is the most common condition in girls (**Figure 43.16**). In cases of imperforate anus with rectovestibular fistula, the rectal orifice is located within the vestibule and outside the hymen. A true rectovaginal fistula is an extremely rare anomaly. Careful inspection of the introitus is important in order to assess for a vaginal septum, or very rarely, distal vaginal atresia.



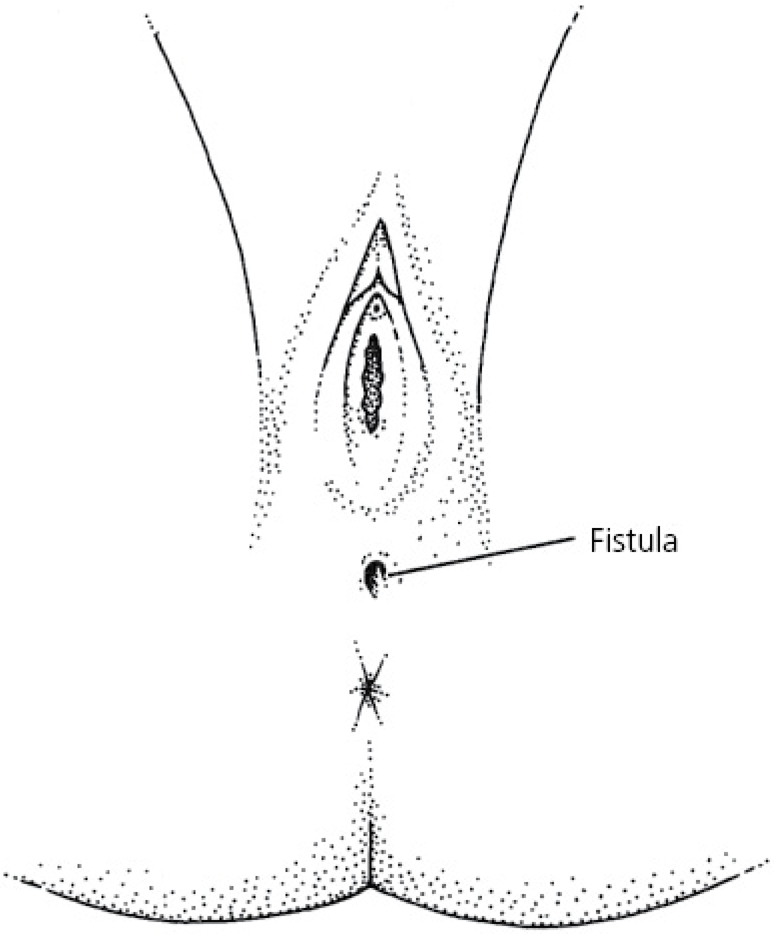
43.16

These patients can undergo a primary repair via a posterior sagittal approach, either in the newborn period or following a period of rectal orifice dilatations provided the surgeon has adequate experience and a meticulous technique. A colostomy followed by the definitive repair is also an acceptable and safe approach.

These fistulas are usually large enough to decompress the gastrointestinal tract. Occasionally, the fistula is too narrow and the patient will have abdominal distension. In these patients, the fistula may first be dilated in order to facilitate emptying of the rectum. The defect is then repaired with a limited posterior sagittal operation.

Patients with rectovestibular fistula are the ones who most often suffer from a failed attempt at a primary repair. Patients with this defect should be expected to be continent after a successful operation. Therefore, an infection and/or dehiscence is particularly problematic as it may damage the continence mechanism and change the final functional prognosis. If this occurs, a redo is often necessary to bring the sphincter muscle back together anterior to the anoplasty.

A rectoperineal fistula is the simplest defect in the spectrum of female malformations (**Figure 43.17**). These patients can be treated with a minimal posterior sagittal anoplasty, without a colostomy, during the neonatal period. The rectal orifice in such patients is too small and usually lies anterior to the center of the sphincter. Repair solves these two problems and additionally provides an adequate length perineal body. Sometimes the small orifice is at the anterior most part of the sphincteric ellipse and only a posterior rectal wall mobilization is needed.



43.17

Most girls with imperforate anus have a fistula (95%). Sometimes, after 20–24 hours of observation, the neonate's abdomen may become distended and there remains no evidence of meconium passing through the genitalia. In such a case, the baby likely has imperforate anus without a fistula. The neonate should undergo a cross-table lateral (cross-fire) radiograph using the same principles discussed for male neonates.

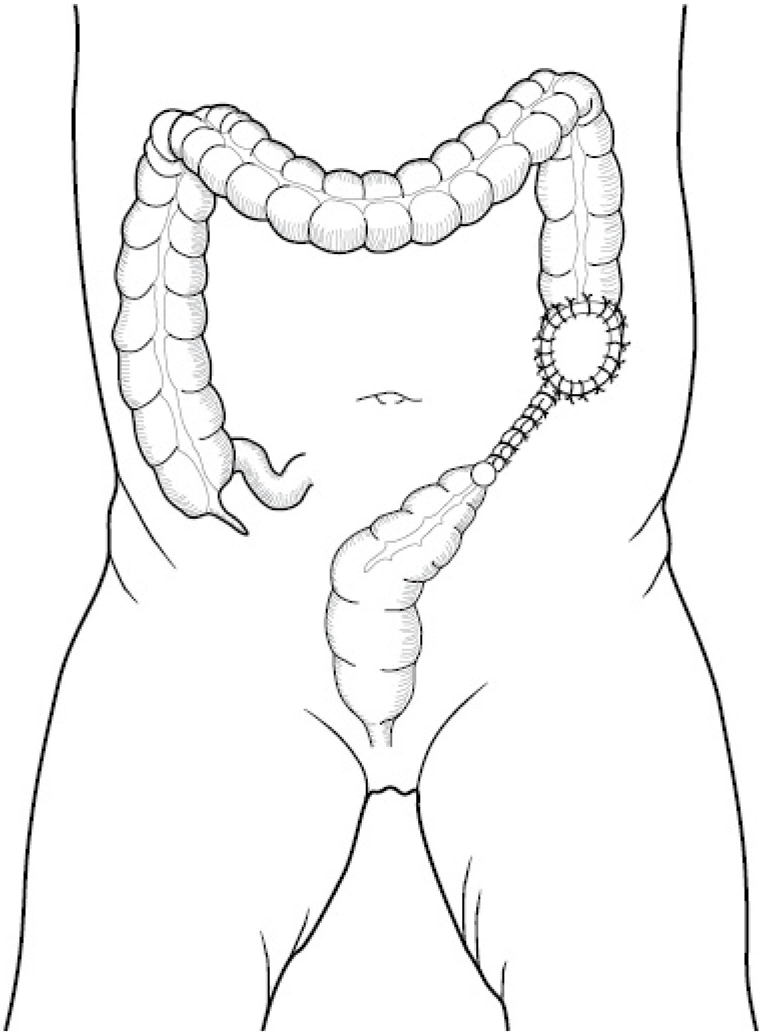
OPERATIONS

Colostomy

A descending colostomy with separated stomas is preferable for the management of ARMs. A well-divided loop (Turnbull) also works well. Transverse colostomies have several disadvantages: The mechanical preparation of the distal colon before the definitive repair is much more difficult and, in the case of a large rectourethral fistula or recto-bladder fistula, the patient can pass urine into the colon, where it remains and is absorbed, which can lead to metabolic acidosis. Also, during the distal colostogram, it is more difficult to distend the distal rectum and thus correctly define the anatomy. Patients with transverse colostomies are more likely to develop a megarectosigmoid. Loop colostomies if they allow the passage of stool from the proximal stoma into the distal intestine, can lead to urinary tract infections and impaction of stool in the distal rectal pouch. They are also more susceptible to prolapse. Prolonged dilatation of the rectal pouch may translate into severe constipation later in life. Colostomy prolapse is more common with colostomies created in a mobile portion of the colon. Prolapse in the proximal limb of a descending colostomy almost never occurs because the proximal stoma is opened immediately distal to where the descending colon is fixed to the left retroperitoneum. A colostomy created too distally in the area of the rectosigmoid colon may interfere with mobilization of the rectum during the pull-through procedure.

During the opening of the colostomy, the distal colon must be irrigated to remove all the meconium, which prevents the formation of a megasigmoid.

A divided colostomy is constructed through a left lower quadrant oblique or transverse incision or using laparoscopy (**Figure 43.18**). The proximal stoma and mucous fistula are exteriorized. The colostomy should be made in the mobile portion of the colon, immediately distal to the descending colon taking advantage of its retroperitoneal attachments, and the mucous fistula is made very small to avoid prolapse. The stomas should be separated enough to allow the use of a stoma bag, which should cover only the functional stoma.



43.18

High-pressure distal colostogram

Before the definitive repair, a distal colostogram is performed. It is the most accurate diagnostic study to define the anatomy of an ARM. Water-soluble contrast medium is instilled into the distal stoma, which fills the distal colon and demonstrates the location of the rectum and the precise site of a rectourinary fistula. Normally, the rectum is surrounded by striated muscle, which keeps it collapsed and prevents filling of the most distal part. This may give the erroneous impression of a very high defect and may prevent demonstration of a rectourinary fistula. To avoid this problem, the contrast medium must be injected with considerable hand-controlled hydrostatic pressure under fluoroscopic or ultrasound guidance. The use of a Foley catheter is recommended; it is passed into the distal stoma, the balloon is inflated (2 cm^3), and it is pulled back to occlude the stoma below the fascia during the injection of the contrast medium. This maneuver permits exertion of enough hydrostatic pressure to overcome the muscle tone of the sphincter muscle mechanism, fill the rectum, and demonstrate a urinary fistula when present.

In cases of rectourethral fistula (prostatic and bulbar), the surgeon knows from the study where to find the rectum. In cases of recto-bladder neck fistula, the surgeon does not expect to find the rectum through the posterior sagittal approach and thus can avoid a blind perineal dissection. The surgeon can properly prepare the patient for laparoscopy or laparotomy to mobilize a higher rectum. The main anatomic questions are whether the distal rectum extends to below the pubo-coccygeal line, whether it is reachable through a posterior sagittal incision, or if laparoscopy and a transabdominal approach is better.

Incision for definitive repair

Using a posterior sagittal approach, the length of the incision will depend on the specific defect. The patient is placed in the prone position with the pelvis elevated. An electric stimulator is used to elicit muscle contraction during the operation as a guide to remain exactly in the midline. The anal dimple should be marked prior to making the incision. An incision that starts at the coccyx and extends anteriorly to the anal sphincter is necessary for rectoprostatic fistulas. Smaller incisions (limited posterior sagittal anorectoplasty) are adequate for defects such as a rectovestibular fistula. Rectoperineal fistulas require a very small posterior sagittal incision (minimal posterior sagittal anoplasty). If laparoscopy is used, a minimal perineal incision is required.

The anatomic relationship of the rectum to genitourinary structures is complex and the separation of the rectum from these structures represents the most risky part of the procedure.

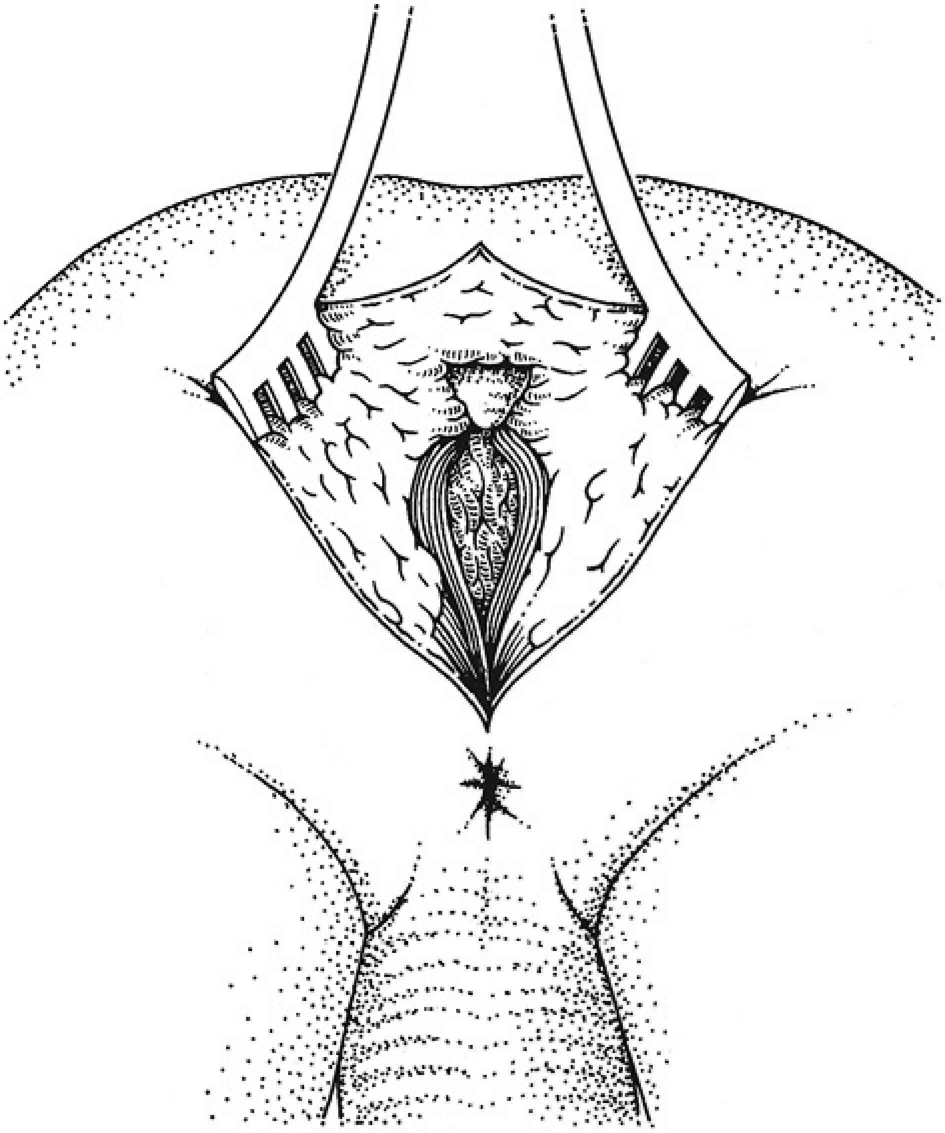
Definitive repair in males

RECTOPERINEAL FISTULA

The repair of these defects consists of a small anoplasty with minimal mobilization of the rectum, sufficient for it to be placed within the limits of the sphincter. The posterior extent of the incision can be the location of the ultimate posterior aspect of the anoplasty. Sometimes only the posterior rectal wall needs to be mobilized completely avoiding an anterior rectal wall dissection. This is a meticulous operation and one that can be done during the neonatal period without a colostomy. The most common complication during the repair of this defect is a urethral injury, which can be avoided by placing a urethral catheter and taking particular care during the dissection of the anterior rectal wall. Dissection adjacent to the urethra causes spongiosum tissue bleeding and is an important clue for the surgeon that they are too close to the urethra.

Patients with this defect have an excellent prognosis. They have problems with bowel control if they undergo an inadequate surgical technique, if they are undertreated for constipation, or if they have significant associated spinal or sacral problems.

The skin is opened through a mid-sagittal incision, and the parasagittal fibers and muscle complex are divided exactly in the midline with a fine-needle cautery (**Figure 43.19**). The fibers of the muscle complex run perpendicular and medial to the parasagittal fibers. The crossing of the muscle complex fibers with the parasagittal fibers represents the anterior and posterior limits of the new anus. These limits can be seen most clearly with the use of an electrical stimulator.

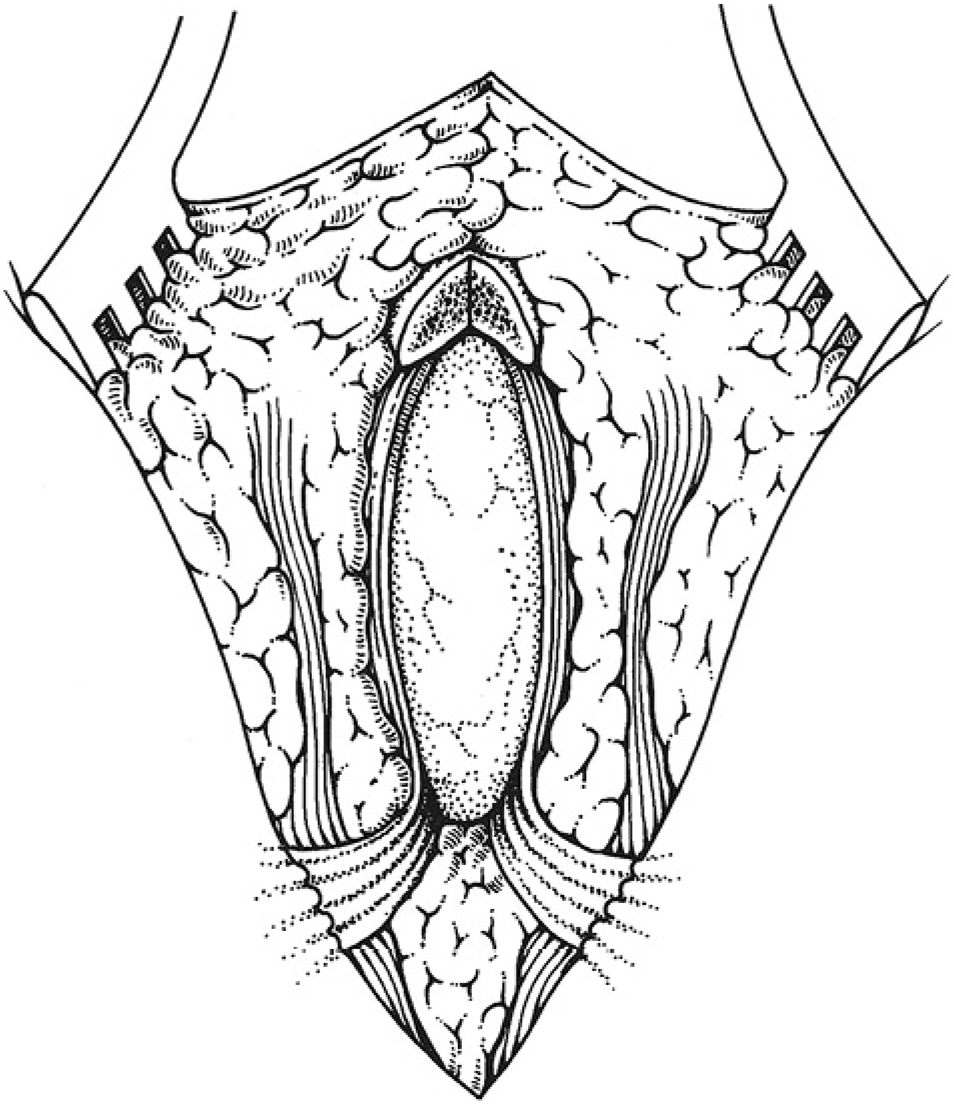


43.19

RECTOURETHRAL FISTULA

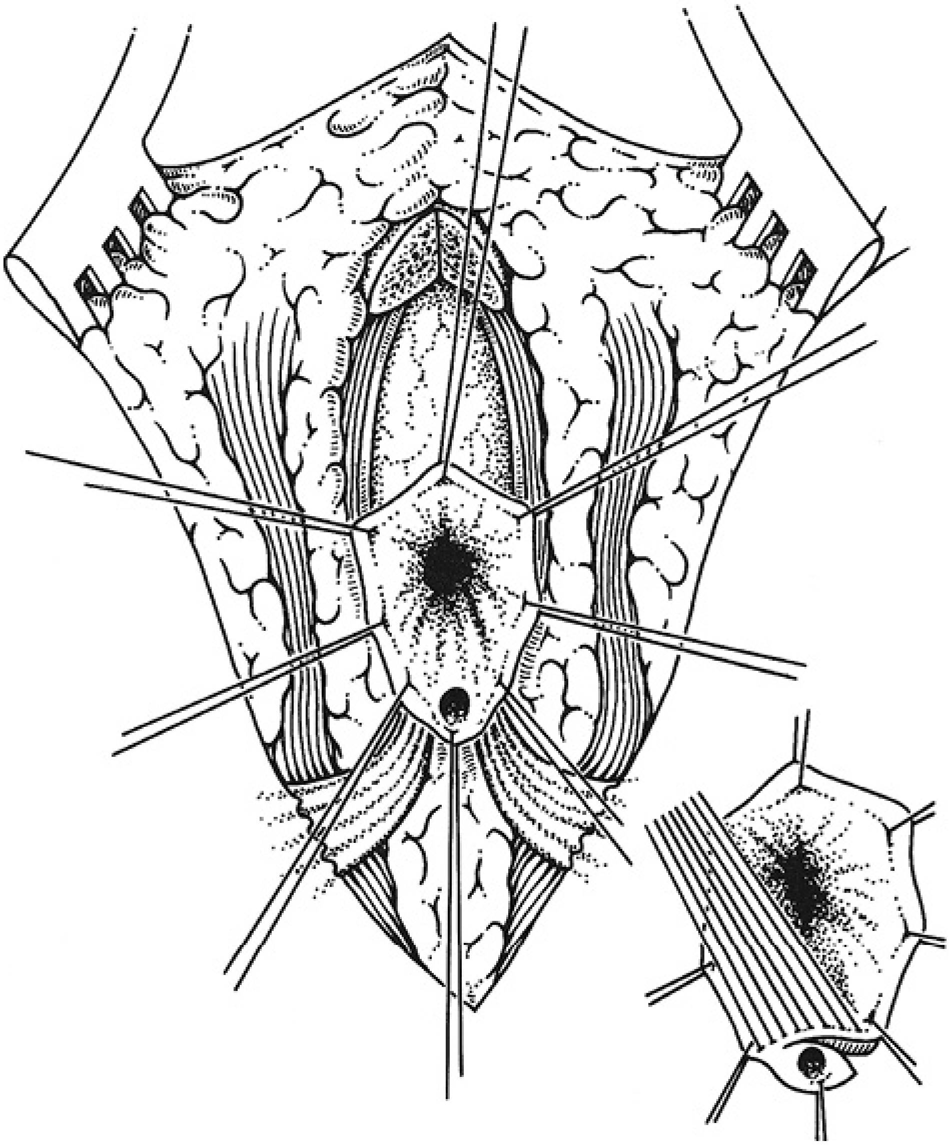
A Foley catheter is inserted through the urethra. To avoid having the catheter enter the rectal fistula, the catheter must be intentionally directed anteriorly. A Coude catheter is a very useful catheter type for this purpose. Occasionally, the catheter must be positioned intraoperatively under direct vision once the fistula is visualized. Cystoscopic insertion of a catheter over a wire is another option.

When all muscle structures have been divided in the midline, the rectum is visible (**Figure 43.20**). In cases of rectourethral bulbar fistulas, the distal rectum is prominent and it almost bulges into the wound. In cases of rectoprostatic fistulas, the rectum is located much higher, just under the coccyx, and is not as prominent. The levator muscles must be separated to expose the rectum at this height. In cases of recto-bladder neck fistulas, the rectum is not visible through this approach, and searching for it risks injuring other midline, whitish structures.



43.20

Two silk sutures are placed in the posterior rectal wall on either side of the midline. The rectum is opened between the sutures and incised distally, exactly in the midline, down to the fistula site. Temporary silk sutures are placed on the lateral and anterior edges of the open posterior rectal wall for traction (**Figure 43.21**). One silk stitch is placed at the location of the fistula to serve as a guide to locate the fistula site so it can be sutured closed after the rectum is separated from the urinary tract.



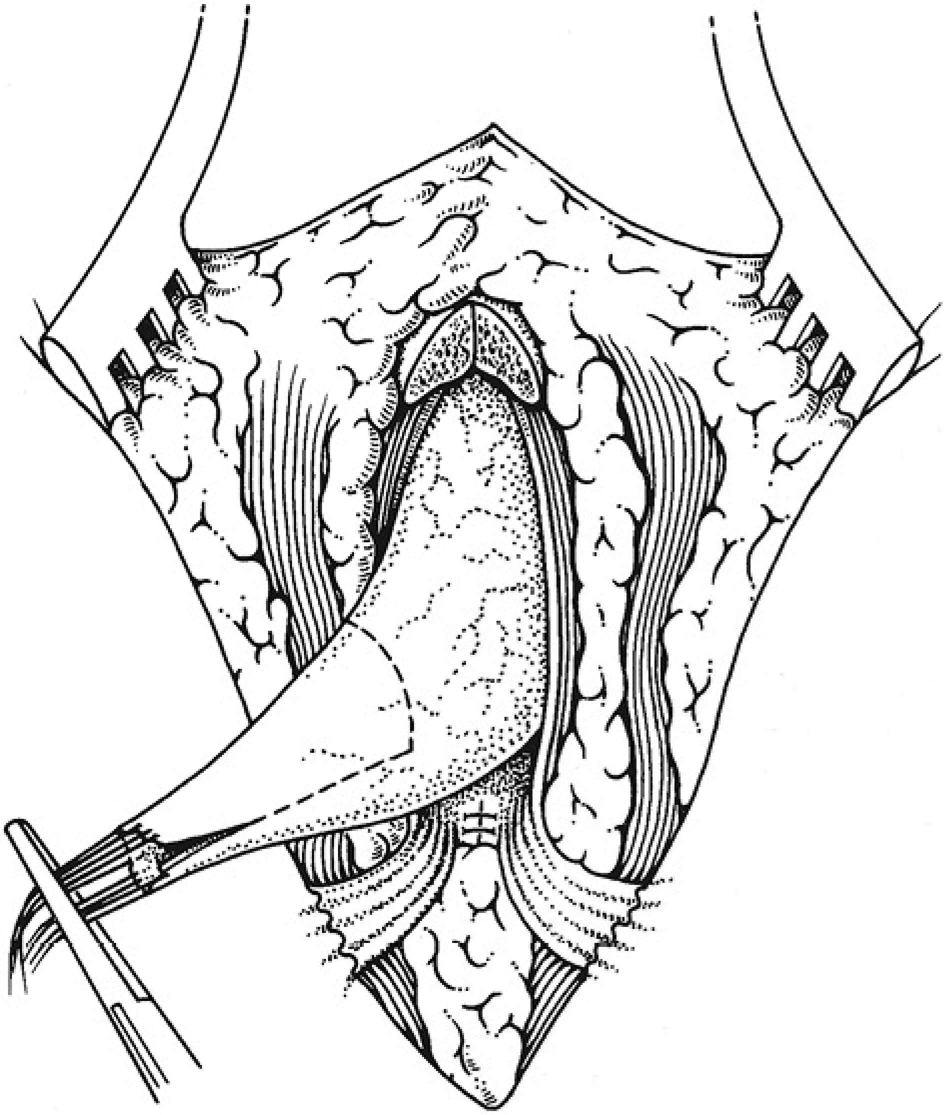
43.21

The anterior rectal wall immediately above the fistula is a thin structure. There is no plane of separation between the rectum and urethra in that area, and one must be created in the common wall. Multiple silk sutures are placed through the rectal mucosa immediately above the fistula in a semicircle. The rectum is then separated from the urethra, creating a submucosal plane for approximately 5–10 mm above the fistula site until the rectum is free from its adherence to the periurethral tissue. The rectum is surrounded by a conspicuous whitish fascia. The dissection must be performed between this fascia and the rectal wall to successfully mobilize the rectum and avoid damage to the innervation of the bladder and genitalia.

During this delicate dissection, it is very helpful to start with the lateral rectum first, very close to the rectal wall and only then go anteriorly, until both dissections (lateral and medial) meet, separating the rectum completely from the urinary tract. A typical areolar plane is then seen between rectum and urethra. Once the rectum is fully separated, a circumferential perirectal dissection is performed to gain enough rectal length to reach the perineum.

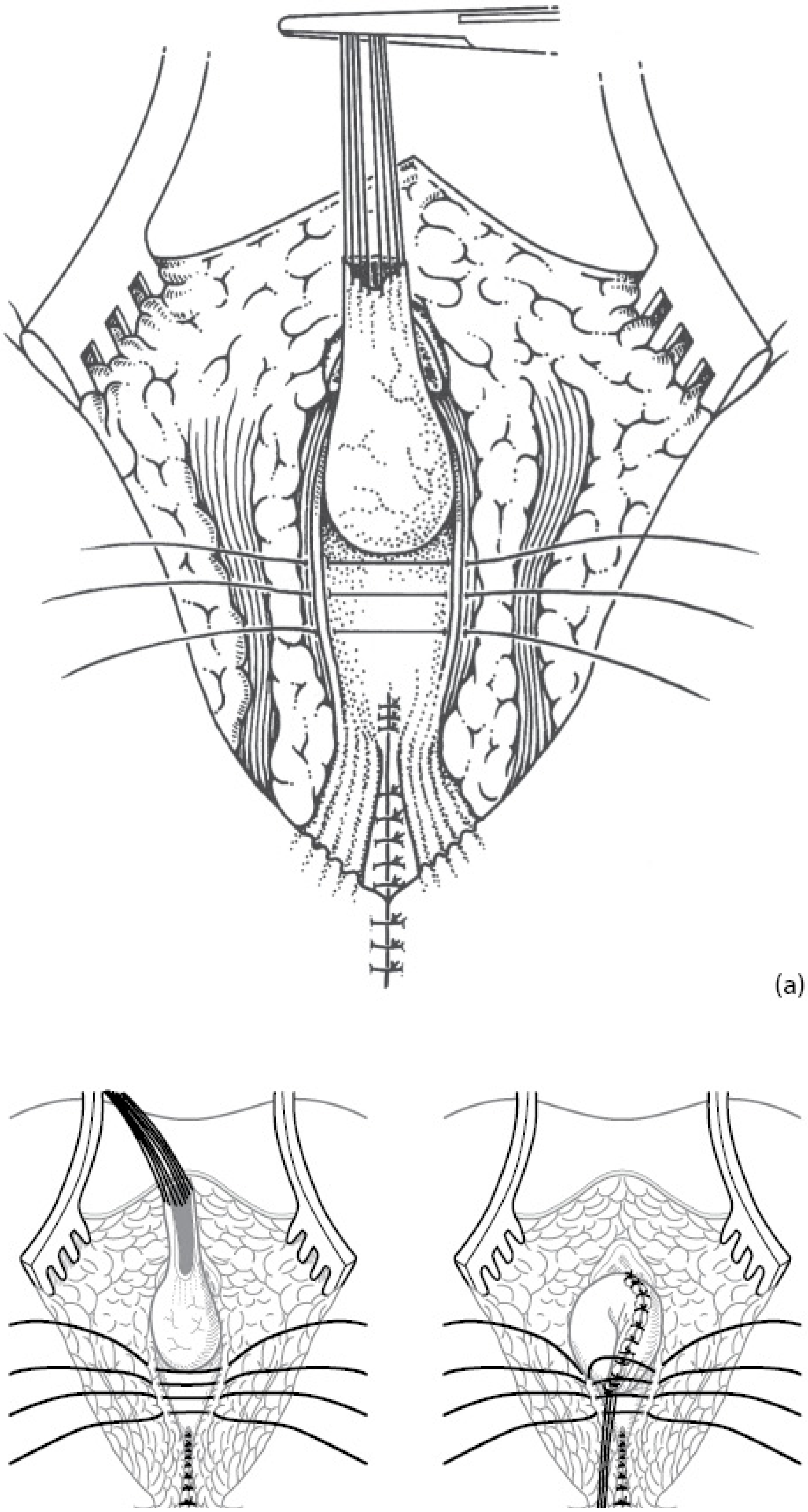
In cases of a fistula opening into the bulbar urethra, the dissection necessary to pull the rectum down to the perineum is minimal, whereas in cases of prostatic fistula the perirectal dissection is considerable. In both cases, enough rectal length must be gained in order to perform a comfortable, tension-free anastomosis between the rectum and the skin. As traction is exerted on the mobilized rectum, some grooves can be seen in the rectal wall, which demonstrate the tissues that hold the rectum. These indentations are bands that must be divided. For a high prostatic fistula, a laparoscopic approach for this mobilization may be helpful, similar to that described for a recto-bladder neck fistula.

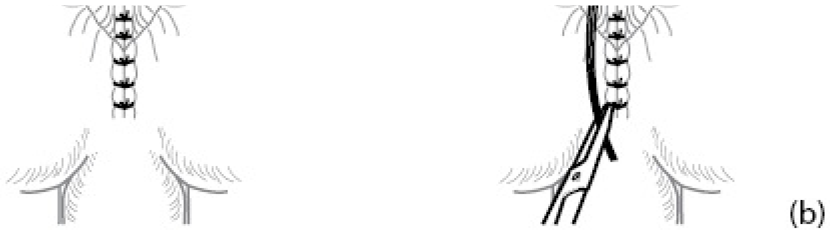
Once the rectum has been fully mobilized, a decision must be made as to whether to taper the rectum. The size of the rectum can be compared to the available space so that its size matches the limits of the sphincter. Rarely necessary, but if needed, the rectum can be tapered by removing part of the posterior wall, and reconstructing it with two layers of interrupted, long-lasting, absorbable sutures (**Figure 43.22**). The tapering must always be done on the posterior rectal wall so that the closed rectourethral fistula is adjacent to normal rectum to avoid creating a fistula. The rectourethral fistula needs to be sutured longitudinally with a single layer of long-term, absorbable sutures.



43.22

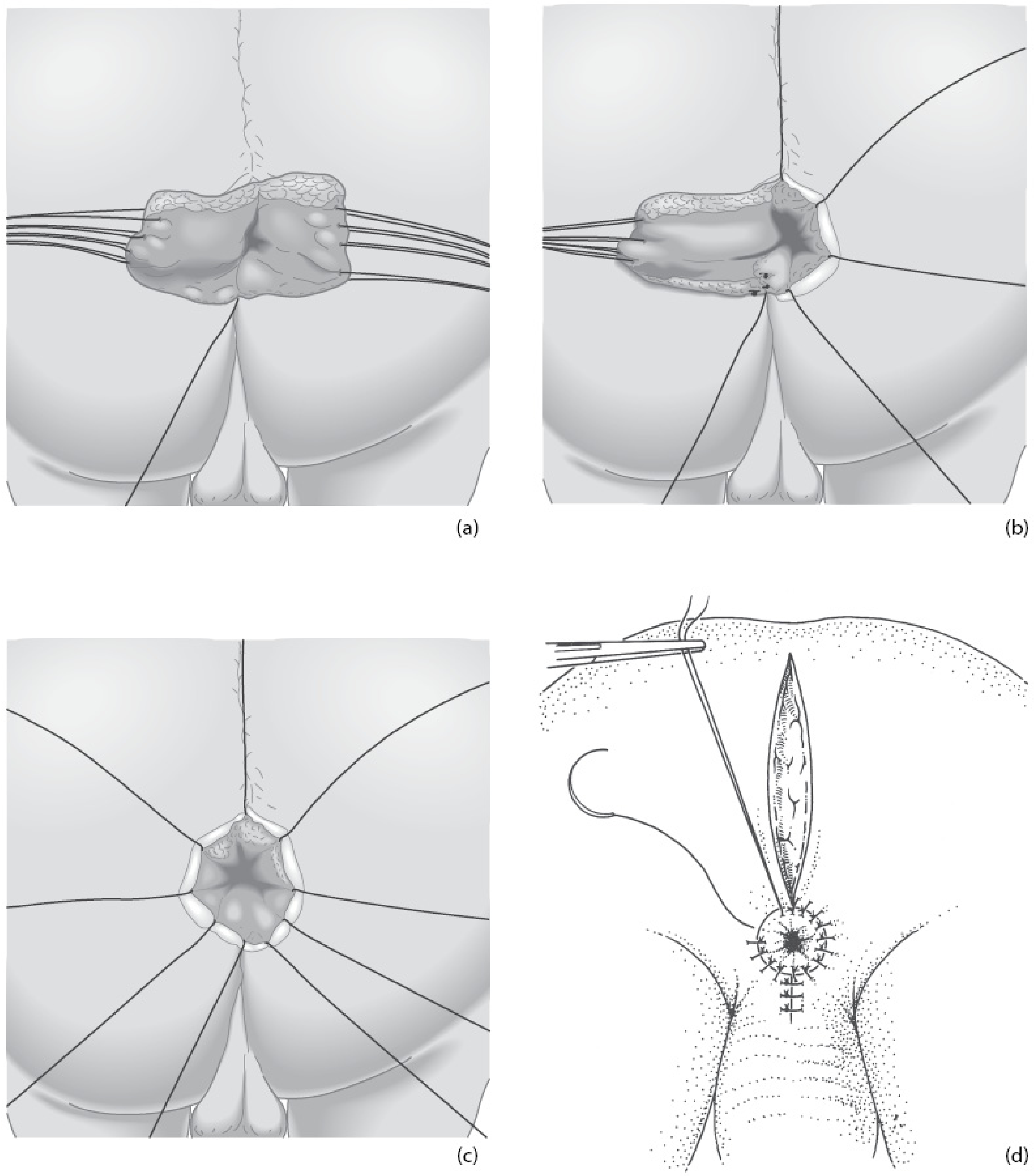
The rectum is placed in front of the levator muscle and within the limits of the muscle complex and external sphincter (**Figure 43.23a**). The electrical stimulator is helpful in identifying the limits of these muscle structures. This location should match the anoplasty marked at the start of the case. In cases in which the incision is extended anteriorly beyond the limits of the sphincter, it is necessary to repair the anterior perineum with interrupted, long-term, absorbable sutures to bring together both anterior limits of the external sphincter (**Figure 43.23a**). Long-term absorbable sutures are placed to reapproximate the posterior edge of the levator muscle. The posterior limit of the muscle complex must also be closed behind the rectum (**Figure 43.23b**). These sutures should include part of the rectal wall in order to anchor it and help to avoid rectal prolapse.


(a)



43.23a,b

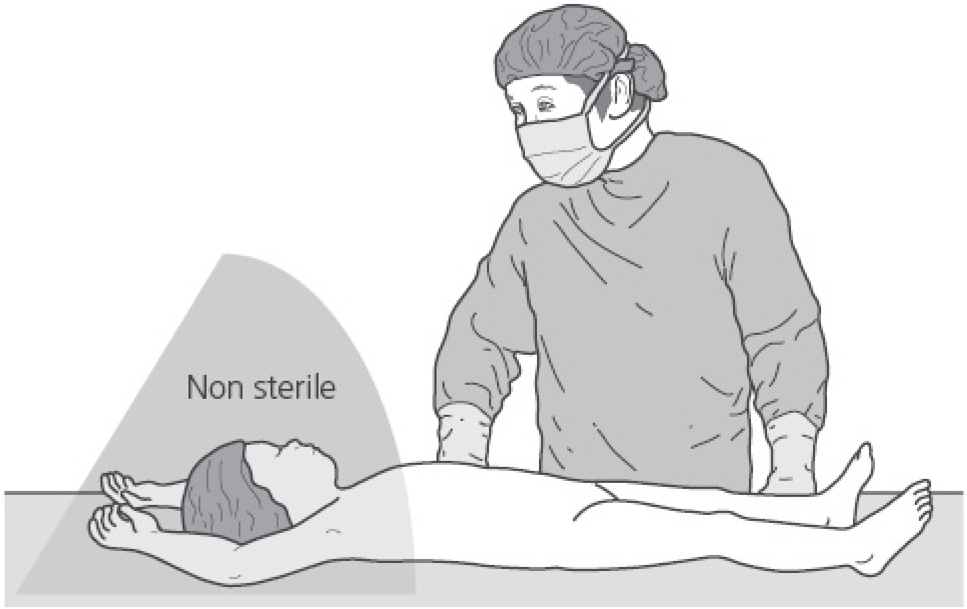
The anoplasty is performed with circumferential interrupted, absorbable sutures. Anoplasty sutures are placed under slight tension, so that once cut, the anus retracts slightly. The posterior sagittal incision is then closed, bringing together corresponding sphincteric structures in the midline (**Figure 43.24a–d**).



43.24a–d

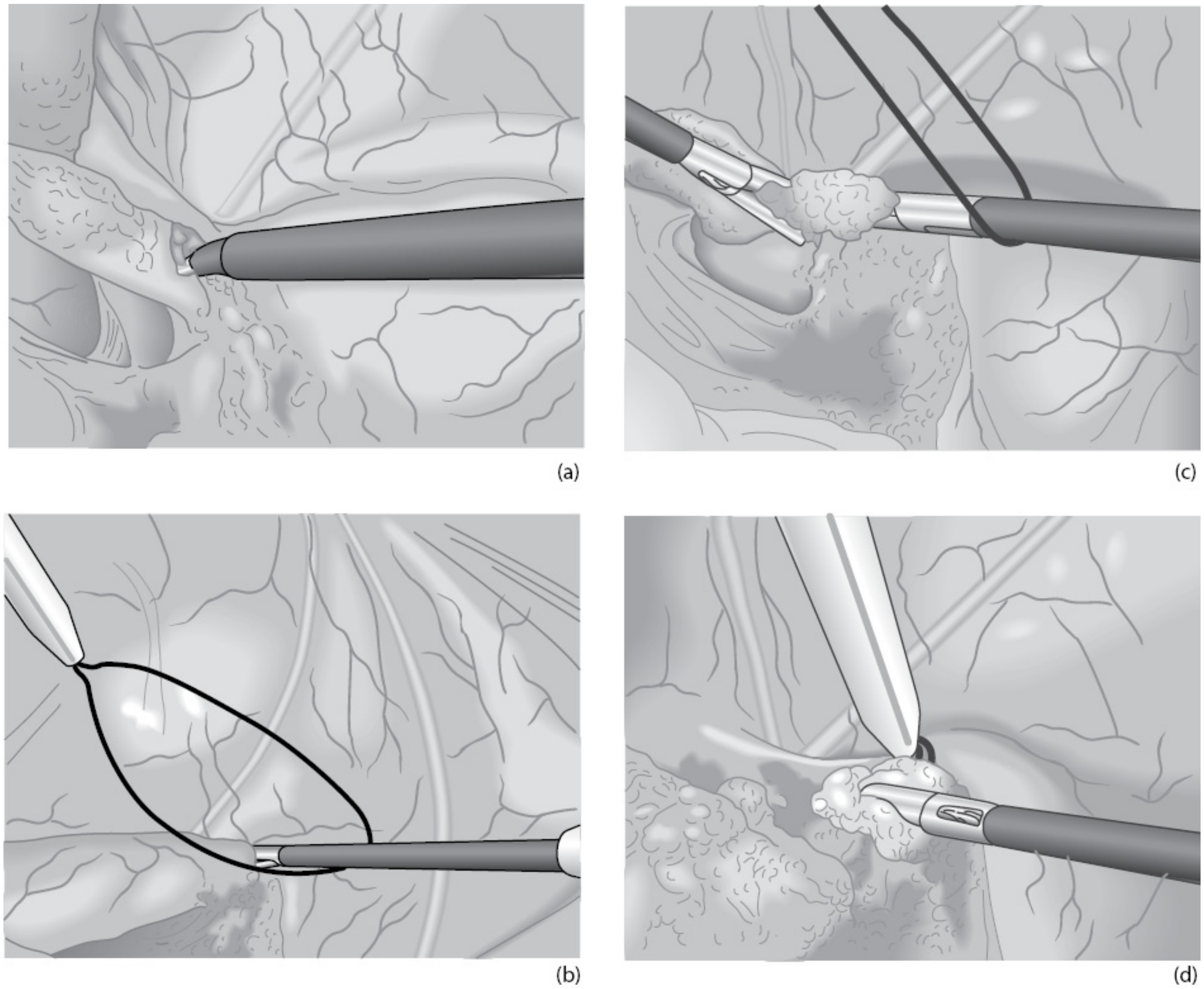
RECTO–BLADDER NECK FISTULA

For the repair of recto-bladder neck fistulas, the entire lower part of the patient’s body from chest down is included in the sterile field, so that the surgeon can work simultaneously in the abdomen and the perineum (**Figure 43.25**).



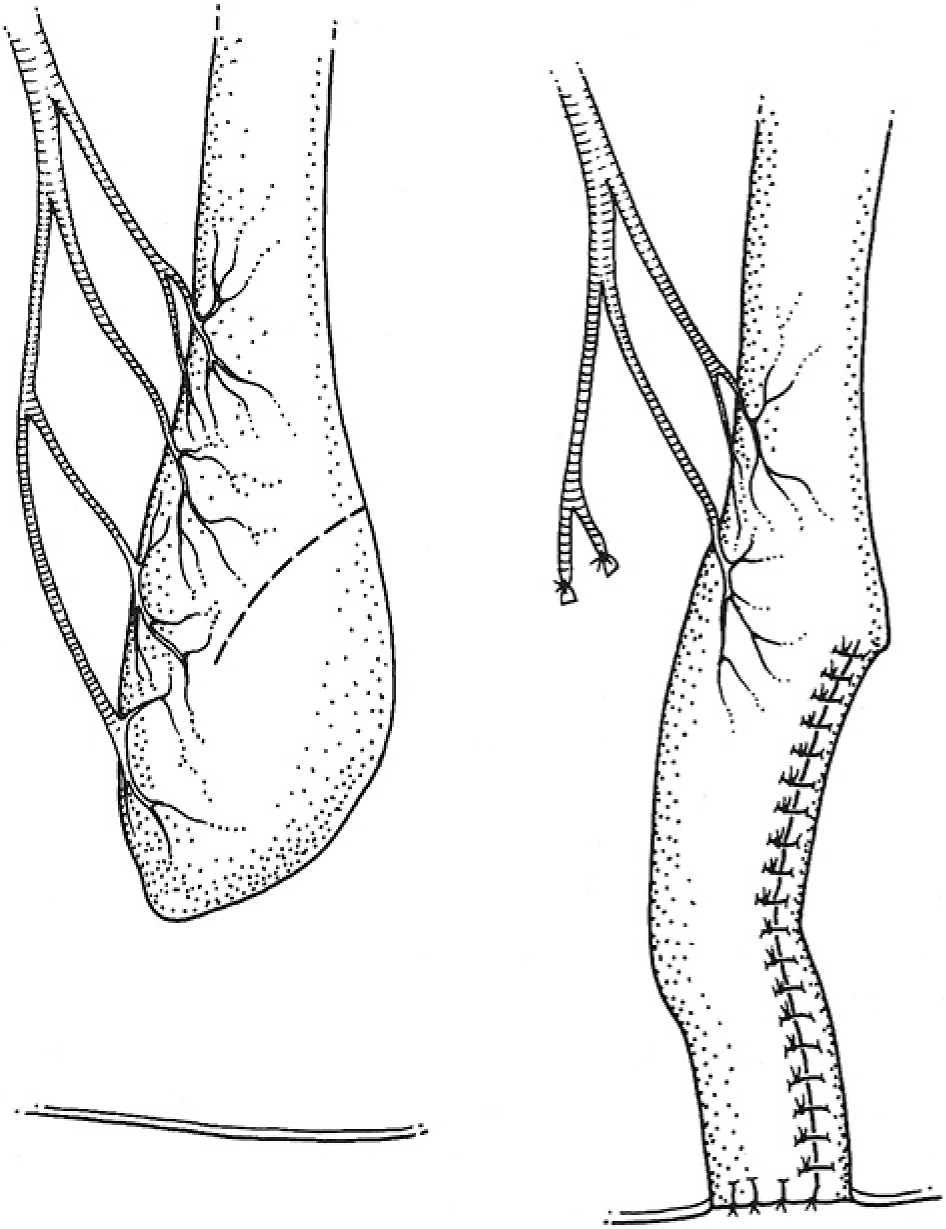
43.25

The abdomen is entered either via laparotomy or laparoscopically, and the distal rectum is mobilized. The peritoneum should be divided around the rectum to create a plane of dissection to be followed distally. The dissection should stay right on the rectal wall. At the point where the rectum becomes narrow, where it communicates with the bladder neck, it should be divided. A useful guide is to dissect the rectum distal enough so that the fistula is narrowed to the width of a 3 mm Maryland. The bladder side of the fistula should be sutured or ligated with an ENDOLOOP® (**Figure 43.26a–d**). In this very high defect, the recto-bladder neck fistula is located approximately 2 cm below the peritoneal reflection, and the rectum communicates with the urinary tract in a T fashion, which means that there is a minimal common wall between the distal part of the rectum and the urinary tract. The surgeon must be careful to avoid damage to the vas deferens, which runs very close to the bowel. With a laparoscopic technique, separation of the rectum from the bladder neck is straightforward. Gaining adequate length, particularly with a high rectum, is challenging, and must be meticulous so as to avoid devascularizing the distal rectum.



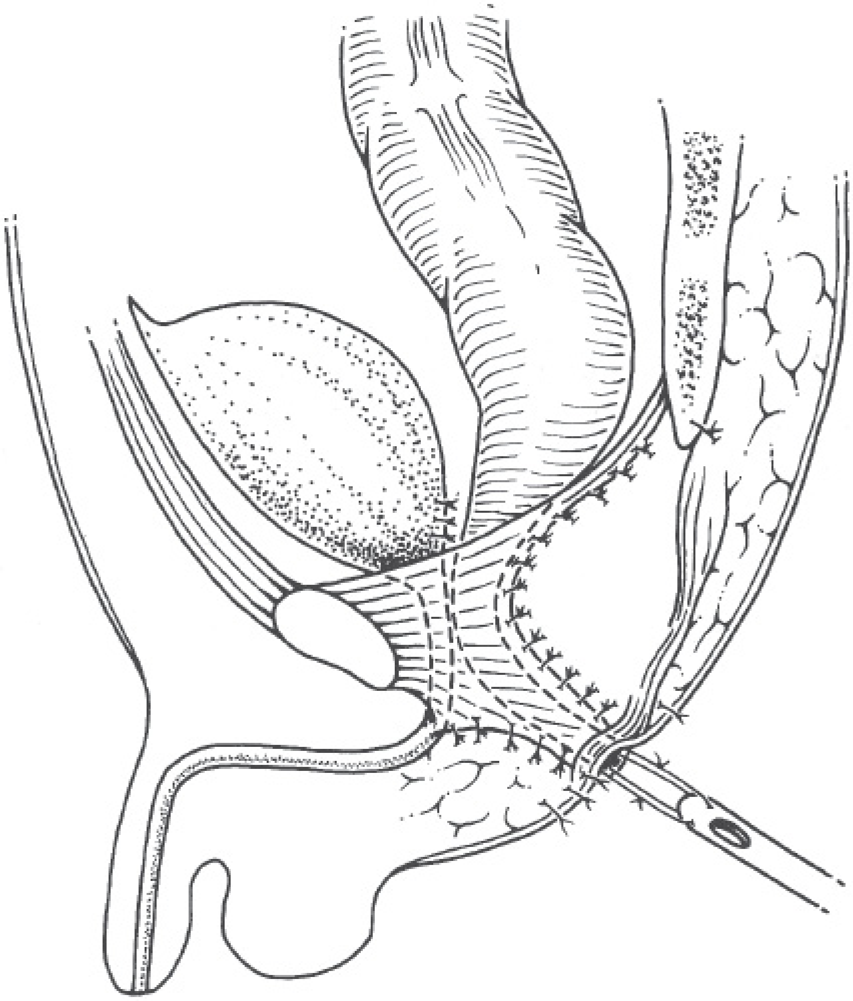
43.26a–d

As this is a very high defect, mobilization of the rectum to reach the perineum requires special maneuvers. Ligation of the inferior mesenteric vessels as high as possible, very close to their origin near the aorta, would mobilize the rectum, but would likely compromise the blood supply of the rectum because the arcades that connect the left colic and sigmoid vessels with the inferior mesenteric ones may have been interrupted at the time of the colostomy creation. Instead, the surgeon must ligate the most distal branches of the inferior mesenteric vessels close to the rectum. If this is done, the more proximal branches of the inferior mesenteric vessels must be left intact to guarantee a good blood supply which runs intramurally to the rectum (**Figure 43.27**).



43.27

If it is necessary to gain extra length, a plasty of the distal dilated portion of the rectum can also be performed. This also functions to taper what is usually a dilated lumen. A combination of these maneuvers usually allows the intestine to reach the perineum, provided the location of the mucous fistula does not interfere with the pull-through of the rectum. This can be anticipated by the distal colostogram, which demonstrates the precise length of intestine available from the colostomy to the end of the rectum. If the colostomy was placed too distal in the sigmoid, the surgeon may have to take down the mucous fistula. In such a case, anastomosis of it to the proximal colostomy with or without a new diversion will allow the distal rectum to reach. The rectum should ideally be preserved as it performs a vital reservoir function. At this point with the legs lifted, a small perineal mid-sagittal incision is performed. The presacral space is dissected (a mosquito clamp dissects gently along the curve of the sacrum) and is visualized through the abdomen. The rectum is pulled through. Great care is taken to avoid urethral injury, as the course of the urethra is directly in front of the course of the future pull-through. Further laparoscopic dissection can be performed while pulling gently on the rectum (Figure 43.28). The rectum is tacked to the muscle complex and an anoplasty is performed as described previously.



43.28

Laparoscopy for this dissection can replace a laparotomy. It provides an excellent view of the peritoneal reflection, the ureters, and the vas deferens; therefore it can be of use in cases of recto-bladder neck fistula where a transabdominal approach is always needed. For high prostatic fistulas, it is also an excellent option.

IMPERFORATE ANUS WITHOUT FISTULA

About 5% of patients have imperforate anus without a fistula. This is the likely defect in a patient with Down syndrome. Most of these patients have a very good sacrum and good muscles. In both males and females, the rectum lies about 2 cm from the perineal skin adjacent to the bulbar level of the urethra. The fact that these patients have no fistula does not necessarily mean that the repair is simpler. The rectum must be carefully separated from the urethra because the two structures have a common wall. The rest of the repair is performed as described for the rectourethral fistula type of defect.

RECTAL ATRESIA AND STENOSIS

These defects are repaired through a posterior sagittal approach. The entire sphincteric mechanism is divided in the midline. The narrowed area of the distal rectum is opened posteriorly. The distal aspect of the anal canal is not mobilized, just opened posteriorly. The dentate line thus converts from a circle to a hemicircle. The posterior rectum in the case of anal stenosis and the full circle of the rectum in the case of rectal atresia is mobilized to reach the anal canal. The mobilized rectum is sutured to the preserved anal canal anteriorly and to anal skin posteriorly to complete the circular anoplasty. The sphincter mechanism posterior to the rectum is reconstructed. A presacral mass must be screened for and is removed during the same operation. If the mass connects to the dura, it should be removed first, with the anal surgery done at a separate setting. The wound is closed following the principles already described. Because the point of maximal tension is at the posterior midline suture, this repair is always covered with a diverting stoma.

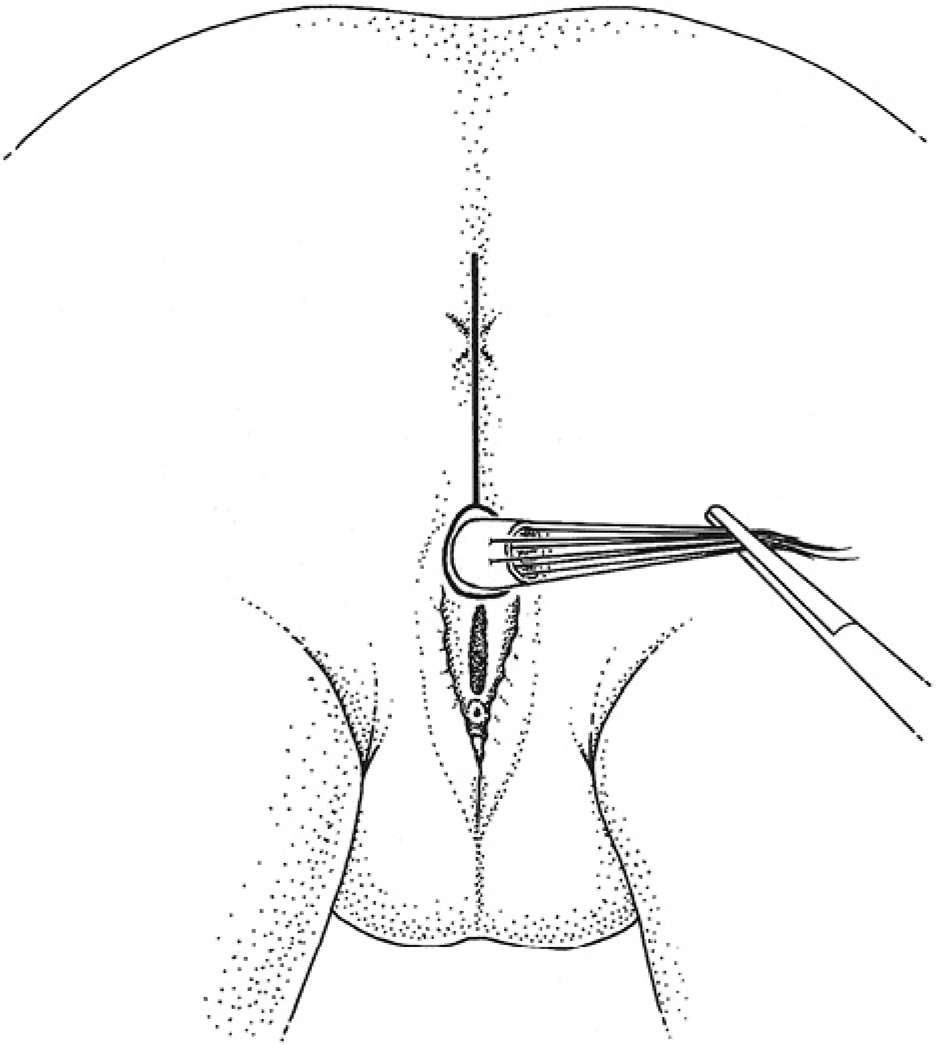
Definitive repair in females

RECTOPERINEAL FISTULA

The treatment of rectoperineal fistula in girls is the same as that discussed for boys, except, of course, that the anterior rectal wall is mobilized off the posterior vagina.

RECTOVESTIBULAR FISTULA

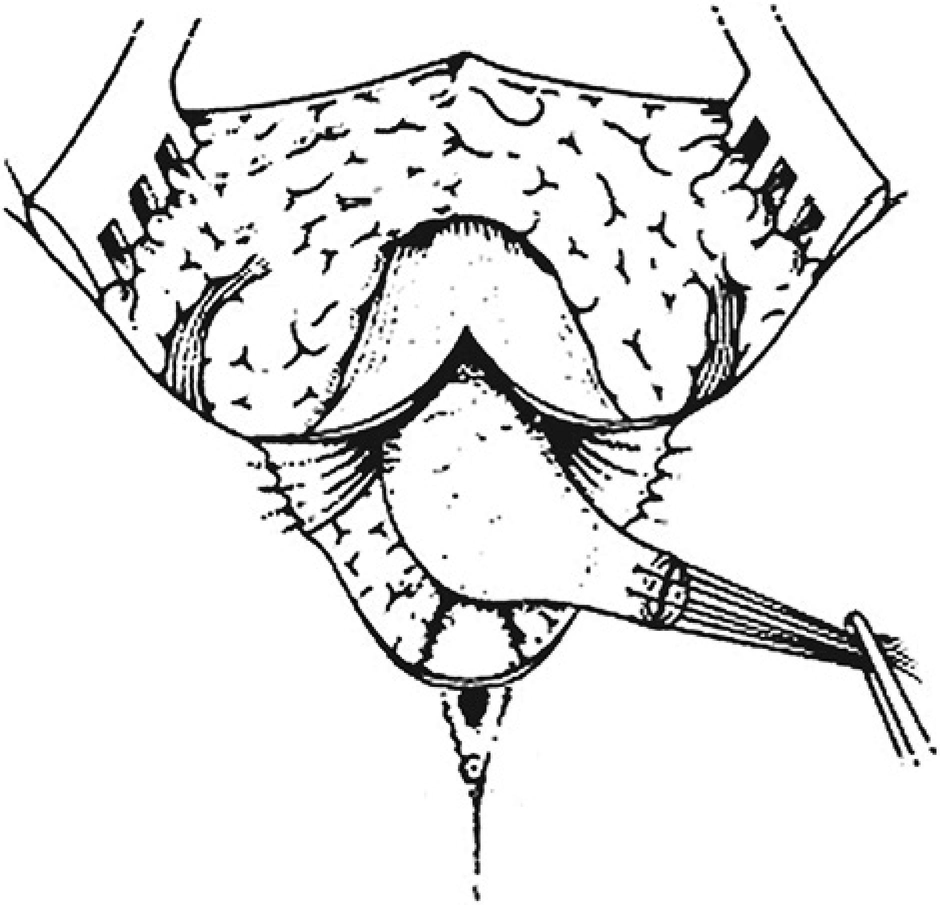
Most surgeons underestimate the complexity of this defect. Multiple sutures are placed at the edge of the fistula in order to exert uniform traction on the rectum to facilitate its dissection (**Figure 43.29**). Sometimes the fistula is deep and hard to see, so the posterior sagittal incision should be made first, and then sutures placed in the fistula orifice. The incision used to repair this defect is shorter than that used to repair rectourethral fistulas in boys. It can extend posteriorly to just at or slightly above the location of the intended posterior aspect of the anoplasty. The dissection continues around the fistula into the vestibule. The sphincteric mechanism is divided in the midline until the rectal wall is located. A characteristic whitish fascia covers the rectum posteriorly and must be dissected off the rectum. This helps to locate the plane of dissection during mobilization of the rectum. Once the rectal wall has been identified, a lateral dissection is performed from the posterior midline, while placing traction on the fistula to make the plane of dissection more obvious. A lateral dissection to clean the rectum facilitates the anterior dissection. It is vital to be adjacent to the rectal wall, and dissect away the thin white fascia that envelops it. The surgeon must be in this key plane in order to mobilize the rectum successfully.



43.29

The most delicate part of the dissection is the anterior rectal wall. The rectum and the vagina share a common wall, which is often very thin. This wall has no plane of separation and the surgeon has to make two walls out of one. This dissection is most easily performed using a fine needle cautery. It is continued up to the point where the rectum and vagina separate and have full-thickness walls. A characteristic areolar tissue between the two walls identifies this point. The most common error in performing this operation is incomplete separation of the vagina and rectum, which may create a tense anastomosis between the rectum and the skin, which can provoke dehiscence of the perineal body.

Once the dissection has been completed, the electrical stimulator is used to determine the limits of the sphincteric mechanism. The limits should be marked at the start of the case before beginning the dissection. The anterior limit of the external sphincter and the anterior edge of the muscle complex are reapproximated as previously described, creating the perineal body (**Figure 43.30**). These sutures are key to allowing for good perineal body healing. The levator muscle is usually not exposed and therefore does not have to be reconstructed. The muscle complex is reconstructed posterior to the rectum with a small bite of rectal wall. The anoplasty is performed as previously described.



43.30

RECTOVAGINAL FISTULA

Imperforate anus with a true rectovaginal fistula is extremely rare. The term is often misused, and patients with a rectovestibular fistula or cloaca are commonly incorrectly described as having a rectovaginal fistula. For a patient with cloaca that is misdiagnosed in this way, the surgeon might repair the rectum but leave the urogenital sinus intact, which would thereafter require a complete reoperation.

A true rectovaginal fistula should be investigated with a distal colostogram. If the fistula is low, it can be repaired posterior sagittally, in essentially the same operation as that described for a rectovestibular fistula, except that it is necessary to dissect much more of the rectum to gain enough length to pull it down to the perineum. Sometimes, if the fistula is high, the rectal dissection can more ideally be done laparoscopically in a very similar way to a recto-bladder neck fistula in a boy. In both such cases, no incision of the perineal body is required. The anterior extent of the incision can end at the anterior limit of the sphincter.

POSTOPERATIVE CARE

Patients generally have a smooth postoperative course. Pain is not a prominent symptom, except in those patients who have undergone a laparotomy.

In cases of rectourethral fistula, the urethral catheter is left in place for 7 days. If the urethral catheter is accidentally dislodged, the patient can be observed for spontaneous voiding, which usually occurs. Attempts to reintroduce a urethral catheter can be dangerous and must be avoided. A suprapubic tube in such a scenario is a better intervention if needed.

IV antibiotics are administered with one pre- and two postoperative doses. Patients with a stoma are usually discharged after 2 days in cases of a posterior approach without a laparotomy or laparoscopy, and after 3–5 days in cases of an abdominal approach. The parents are instructed to keep the incision clean and not to wipe. In children with no stoma, the wound is observed for 5 days while on clear liquids only and then the diet can be advanced if good healing is confirmed.

Two weeks after the operations, anal dilatations may be started but, in many cases, are not necessary. On the first occasion, a dilator that fits loosely into the anus is used to instruct the parents, and the size of the dilator increased until the rectum reaches the desired size, which depends on the patient's age (**Table 43.3**). This process, if undertaken, usually takes approximately 8 weeks. Once the desired size is reached, the colostomy can be closed.

Table 43.3 **Size of dilator required in different age groups**

Age group	Hegar dilator size
1–4 months	12
4–8 months	13
8–12 months	14
1–3 years	15
3–12 years	16
Over 12 years	17

After the colostomy is closed, patients often suffer from diaper rash as a consequence of multiple bowel movements. The number of bowel movements eventually decreases, usually within several weeks, and patients should work toward a good bowel movement pattern, which would be one to two well-formed stools per day. This is achieved with a combination of diet, a Senna-based laxative, and water-soluble fiber. The final functional result of whether a child has voluntary bowel movements can be predicted based on the precise anatomic diagnosis and the status of the sacrum and spine, called the ARM Continence Index (see **Table 43.2**).

FUNCTIONAL EXPECTATIONS

Deficiencies in sensation

Except for patients with rectal atresia and stenosis, patients with ARM are born without an anal canal. This means that they do not have the exquisite sensation that normally resides in this anatomic area. Most patients, however, still preserve a vague sensation called proprioception, generated from distension of the rectum, and therefore stretching of the voluntary muscles around it. Liquid stools, which do not distend the rectum, cannot be felt.

Sphincteric mechanism

ARMs represent a spectrum and most patients have a sphincteric mechanism with different degrees of development which varies from almost non-existent muscles to a nearly normal sphincteric mechanism. Therefore, most of these patients have a certain capacity to hold stool inside the rectum.

Rectosigmoid motility

Most patients with ARMs suffer from abnormal rectosigmoid motility. Patients who have undergone a surgical procedure in which the rectosigmoid colon was removed, such as an abdomino-perineal pull-through, do not have a rectum, but have a segment of sigmoid or descending colon pulled down to the perineum. They have a tendency to pass loose stool, which is hard to manage with suboptimal sphincters and anal canal sensation.

On the other hand, patients who have undergone repair in which the rectosigmoid colon was preserved (e.g. posterior sagittal anorectoplasty, or an anoplasty) have a fecal reservoir that is large and floppy, which leads to varying degrees of constipation. Mild cases of constipation can be treated very effectively with laxatives. Severe cases of constipation may lead to fecal impaction, soiling, and therefore overflow pseudo-incontinence. This constipation seems to be more severe in patients with lower defects.

COMPLICATIONS

Wound infection

Wound infections and mild dehiscences of the posterior sagittal incision and perineal body can occur. The infection usually affects only the skin and subcutaneous tissue, sometimes can be resutured, and will heal with local care. If the perineal body disrupts, it may heal in with no sphincter muscle anterior to the anus, and this often requires a redo operation.

Anal strictures

A severe anal stricture usually requires a secondary operation. A clear correlation exists with intraoperative devascularization of the distal rectum or excessive tension on the anoplasty.

If the protocol of dilatations is not used, sometimes a skin level anal stricture (1–2 mm) can occur. Such a stricture is only a ring-like fibrous band at the mucocutaneous junction, which is easily treated using a Heineke-Mikulicz anoplasty. This could be done at the time of colostomy closure. This situation is different from a long, narrow stricture secondary to ischemia which needs a redo.

Rectal prolapse

Outpouching of the rectal tissue is often due to poor pelvic musculature, but it can also occur with inadequate tacking of the posterior rectal wall to the posterior edge of the muscle complex. A trim of the excess of tissue can be performed, and this can be done as an ambulatory procedure.

Constipation

This is the most common functional disorder observed in patients with ARMs, particularly prevalent in lower malformations (rectoperineal, bulbar, and vestibular fistulas). It should be aggressively managed with stimulant laxatives combined with a water-soluble fiber.

Neurogenic bladder

Difficulty voiding after a posterior sagittal approach should occur only in patients with an abnormal sacrum, tethered cord, or myelomeningocele, in whom the presence of a neurogenic bladder can be expected.

Neurogenic bladder following a posterior sagittal approach in patients with favorable anatomy can occur due to nerve damage during the rectal dissection, if the surgeon does not follow the principles of the posterior sagittal approach and dissects around the rectum too widely.

Medical management of fecal incontinence

Approximately 25% of patients with ARM suffer from soiling, despite optimal anatomic surgical reconstruction. There is clearly a significant functional component to their continence potential that relates to their type of malformation, and the quality of their sacrum and spine. For these patients, a program of bowel management consisting of training the parents and children to clean the colon once a day with the use of enemas and to avoid bowel movements between or by adhering to a specific diet, and sometimes medication, is very valuable. The bowel management program allows the patient to remain clean all day and be in normal underwear.

There are two patterns:

- *Constipated.* These patients have undergone a procedure in which the rectum was preserved, as in anoplasties or a posterior sagittal anorectoplasty, and tend to suffer from constipation in addition to their fecal incontinence. A contrast study shows a megarectosigmoid. Management consists of enemas with volumes of fluids large enough to clean a large rectosigmoid colon (400–600 mL of saline usually with added glycerin, and/or a gentle soap). It is not usually necessary to use any special diet or medication, because the constipation helps the patient to remain clean between enemas.
- *Patients with a tendency to have loose stool.* This group of patients has undergone a type of procedure in which their original rectosigmoid colon

was resected, as with an abdominoperineal pull-through. They have a tendency to have constant passage of liquid stools. A contrast enema shows that the colon runs straight from the splenic flexure down to the anus, and colonic haustrations are apparent all the way down to the perineum, evidence of the loss of the rectum. They almost never suffer from constipation. Management consists of a small enema, a strict constipating diet, and agents that slow colonic motility, such as loperamide pectin, and water-soluble fiber. Patients in whom colonic motility fails to slow are the rare candidates for a permanent colostomy.

POTENTIAL FOR BOWEL CONTROL

Patients born with a favorable type of defect (rectovestibular fistula, rectoperineal fistula, rectourethral bulbar fistula), a good sacrum, a normal spine, and a good sphincteric mechanism, and who underwent an operation that placed the rectum in the correct position, should achieve normal bowel control. In addition, these patients should be able to develop a good bowel movement pattern (one to two well-formed stools per day). They can undergo potty training to have voluntary bowel movements. Often they need help with laxatives to treat constipation.

CANDIDATES FOR A REOPERATION

Patients who underwent an operation that placed the rectum in the wrong place or left it strictured or prolapsed will benefit from a reoperation. Improving the rectal anatomy and ensuring it is the right size and within the limits of the sphincter mechanism may improve the functional result.

CANDIDATES FOR A SIGMOID RESECTION

There is a subgroup of patients who were born with a defect that has a good prognosis and who underwent a technically good operation but suffer from severe constipation and a megasigmoid colon. Often such patients' constipation was not aggressively managed from infancy until toddler age. They are incapable of emptying the rectosigmoid colon and suffer from chronic soiling and overflow pseudo-incontinence. Prior to considering a sigmoid resection, a laxative test is performed. The test is carried out over several days. First, the colon is disimpacted with enemas. This process is radiologically monitored. Then the laxative requirement is determined by trial and error. Once the right dose is reached, as demonstrated by an abdominal radiograph that shows a clean colon, the patient's ability to have bowel control can be determined. If the patient is continent, but in order to remain clean requires an enormous dose of stimulant laxatives, a sigmoid resection, preserving the rectum and creating an anastomosis between the descending colon and the rectum above the peritoneal reflection, can reduce that laxative requirement dramatically.

VIDEOS

OPS_8e_video_43.1 Repair female imperforate anus–no fistula

OPS_8e_video_43.2 Cloaca repair with total urogenital mobilization (TUM)

OPS_8e_video_43.3 Repair imperforate anus male with rectourethral bulbar fistula

FURTHER READING

Falcone RA, Levitt MA, Peña A, Bates MD. Increased heritability of certain types of anorectal malformations. *J Pediatr Surg* 2007; 42: 124–8.

Halleran DA, Ahmad H, Vianova-Sanchez A, et al. Assessing the benefit of reoperations in patients who suffer from fecal incontinence after repair of their anorectal malformation. *J Ped Surg* (in press) https://doi.org/10.1016/j//jpedsurg.2020.06.011

Hong AR, Acuña MF, Peña A, et al. Urological injuries associated with the repair of anorectal malformations in male patients. *J Pediatr Surg* 2002; 37: 339–44.

Levitt MA, Peña A. Anorectal malformations. *Orphanet J Rare Dis* 2007; 2: 33.

Shaul DB, Harrison EA. Classification of anorectal malformations – initial approach, diagnostic tests, and colostomy. *Semin Pediatr Surg* 1997; 6: 187–95.

44

Cloaca

ANDREA BISCHOFF AND ALBERTO PEÑA

HISTORY

A persistent cloaca (from the Latin word *cloaca* meaning sewer or drain) is a malformation in which the rectum, vagina, and urethra are fused into a single common channel. This may have a variable length from 1 to 10 cm and is considered one of the most formidable challenges in pediatric surgery. Prior to 1982, there were few reports, other than anecdotal ones, on the treatment of this condition. Even then, these largely concentrated on the repair of the rectal component only and many patients born with a cloaca frequently ended up having a hysterectomy. Most surgical approaches were limited in their exposure and thus could not clearly define the complex anatomy of the defect.

W. Hardy Hendren III at the Boston Children's Hospital contributed much on the secondary reconstruction of these cases and the surgical repair of the associated urologic malformations. He emphasized a global approach to repair of the entire anomaly, but with a focus on urology.

The senior author used the posterior sagittal approach to repair a cloaca in an operation performed in Monterrey, Mexico, in 1982. This demonstrated clearly the delicate, precise anatomic characteristics of the condition. The rectum was separated from the vagina and pulled down to the perineum, the vagina was separated from the urethra and pulled down to the perineum, and what used to be the common channel was reconstructed as a neourethra. The procedure was named and is still known as posterior sagittal anorectovaginourethroplasty (PSARVUP). This patient is now an adult, is urinary and fecally continent, sexually active, and has already delivered a baby by Caesarian section. In retrospect, after learning many lessons from more than 580 operations, we now realize that this first case belongs on the "good" side of the spectrum. This clinical variation in cloacal functional anatomy and physiology may require implementation of a range of technical surgical maneuvers.

PRINCIPLES AND JUSTIFICATION

Incidence

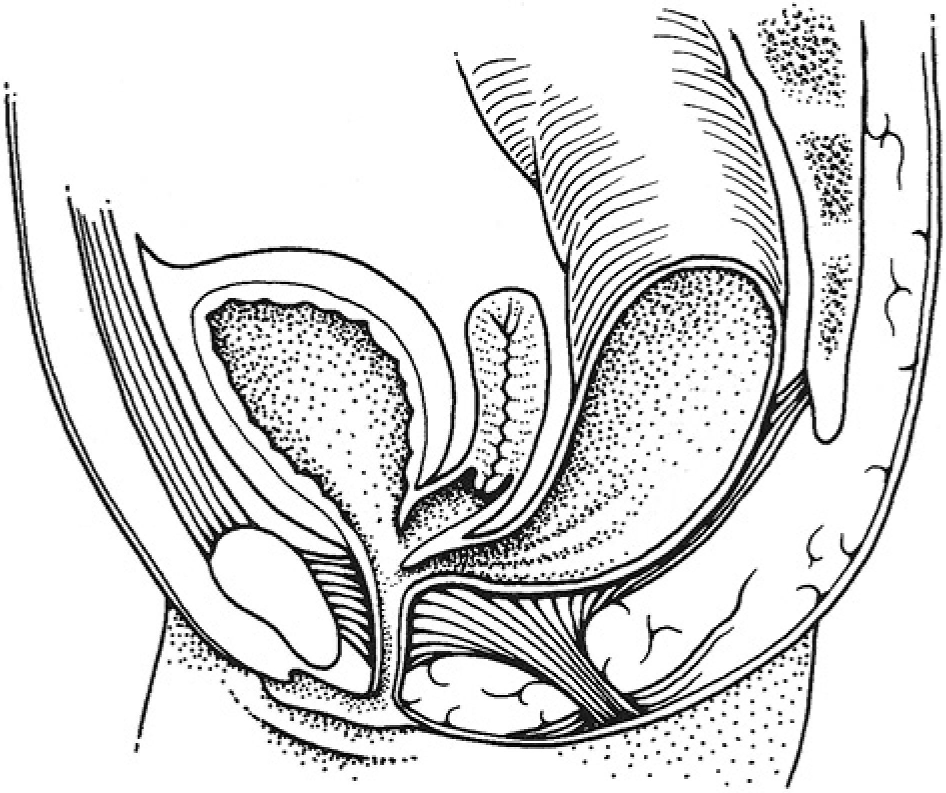
Cloacal anomalies occur exclusively in females and have an incidence of about 1 in 20 000.

In the past, persistent cloaca was considered to be a very unusual defect. However, before 1982 there appeared to be a higher incidence of congenital "rectovaginal fistulas" reported in the literature. In retrospect, it seems likely that many patients with persistent cloaca were erroneously thought to have a rectovaginal fistula. Real rectovaginal fistula is an extremely unusual, perhaps even non-existent, defect. We have seen many patients who have been operated on with the erroneous diagnosis of a "rectovaginal fistula" and who have been left with a persistent urogenital sinus.

The goals of treatment of cloaca include reconstruction of the urethra, vagina, and rectum, and the achievement of bowel control, urinary control, and sexual function.

Classification

The length of the common channel has important therapeutic and prognostic implications (**Figure 44.1**). Based on the long-term follow-up of our patients, we now divide the cases into two groups. This classification is rather simplistic, but it has important therapeutic and prognostic implications (**Table 44.1**).



44.1

Table 44.1 Short versus long cloaca*

	Group A (n = 260)	Group B (n = 220)
Common channel	Short, <3 cm	Long, >3 cm
Type of operation	Only posterior sagittal	Posterior sagittal and laparotomy
Hospitalization	48 hours	Several days
Associated urological defects (%)	59	91
Incidence in our series (%)	62	38
Voluntary bowel movements (%)	68	44
Urinary continence (%)	72	28
Average number of operations[a]	9	18
Intraoperative decision making	Relatively easy, reproducible operation	Complex, delicate and technically demanding[b]

[a] Including orthopedic, urologic, cardiac, and general.

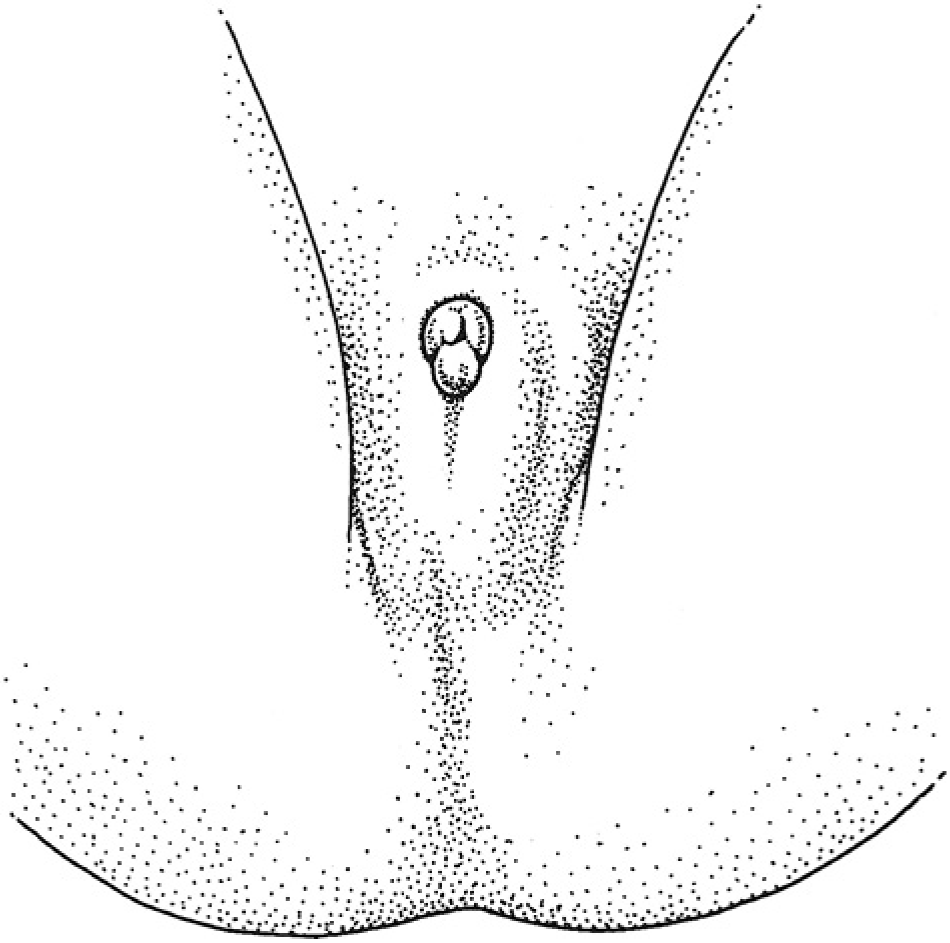
[b] Bladder/vagina separation with or without the following procedures: Ureteral catheter, ureteral reimplantation, vesicostomy, cystostomy, bladder neck reconstruction or closure, vaginal switch, vaginal replacement (rectum, colon, small bowel).

*Information not available for 100 patients (reoperations)

- *Common channel <3 cm:* The most accurate way to measure the length of the common channel is with a cystoscope. When the common channel is <3 cm, the malformation can be repaired by a posterior sagittal approach without laparotomy. These patients usually have a well-developed sacrum, good sphincters, and a good functional prognosis.
- *Common channel >3 cm:* Most cases in which the common channel is >3 cm will require an additional transabdominal approach and operation should ideally only be undertaken by those with significant experience in the management of these malformations. The functional prognosis for both bowel and urinary control may be less than optimal.

DIAGNOSIS

Careful inspection of the perineum, including a separation of the labia minora, discloses a single perineal orifice, which is pathognomonic of a cloaca (**Figure 44.2**). Sometimes there is a palpable lower abdominal mass that represents a distended vagina (hydrocolpos). Failure to recognize the presence of a cloaca in a neonate may be dangerous, as more than 90% have important associated urologic problems.

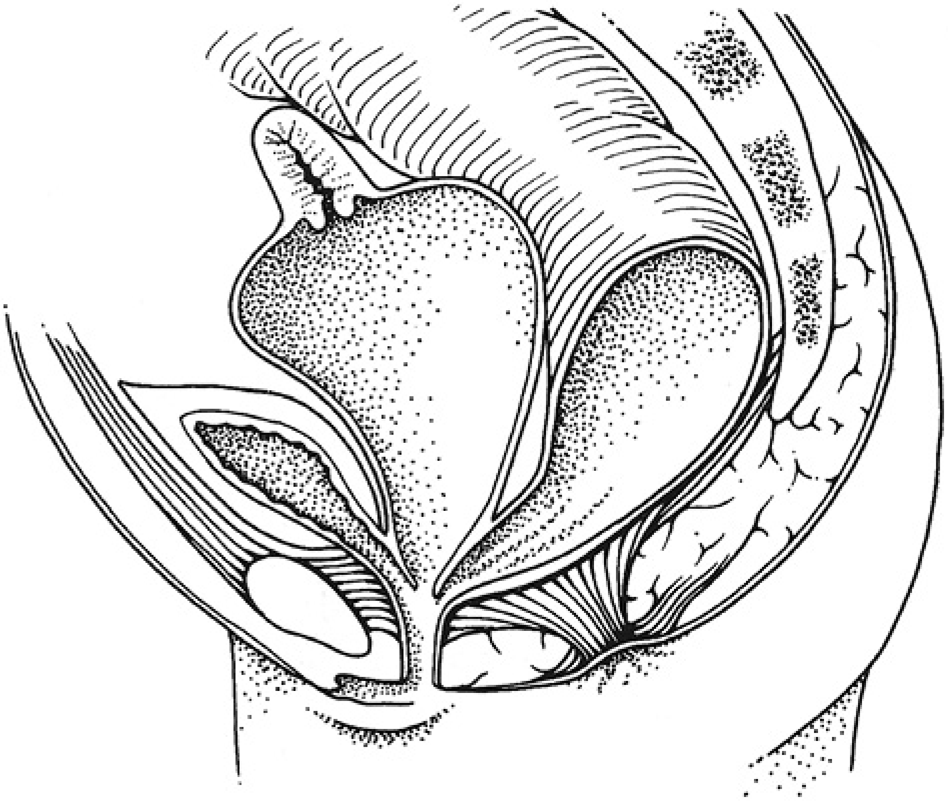


44.2

MANAGEMENT OF CLOACAL MALFORMATIONS DURING THE NEONATAL PERIOD

Once the clinical diagnosis of a cloaca has been established, the next step is to perform an emergency urologic evaluation, which includes an abdominal and pelvic ultrasound (US) to exclude hydronephrosis, hydroureter, and/or hydrocolpos.

The vagina is abnormally distended in 30% of cases, and full of urine and mucus (hydrocolpos) (**Figure 44.3**). This may compress the trigone of the bladder, producing an extrinsic ureterovesical obstruction, megaureters, and hydronephrosis. The most common error at this stage is to perform only a colostomy in a patient with severe obstructive uropathy, as this can lead to acidosis and urinary sepsis. The dilated vagina can also become infected. This condition is called pyocolpos, and it may lead to vaginal perforation and peritonitis. Such a large vagina may ultimately represent a technical advantage at the time of the main repair, because having more vaginal tissue will facilitate its reconstruction.

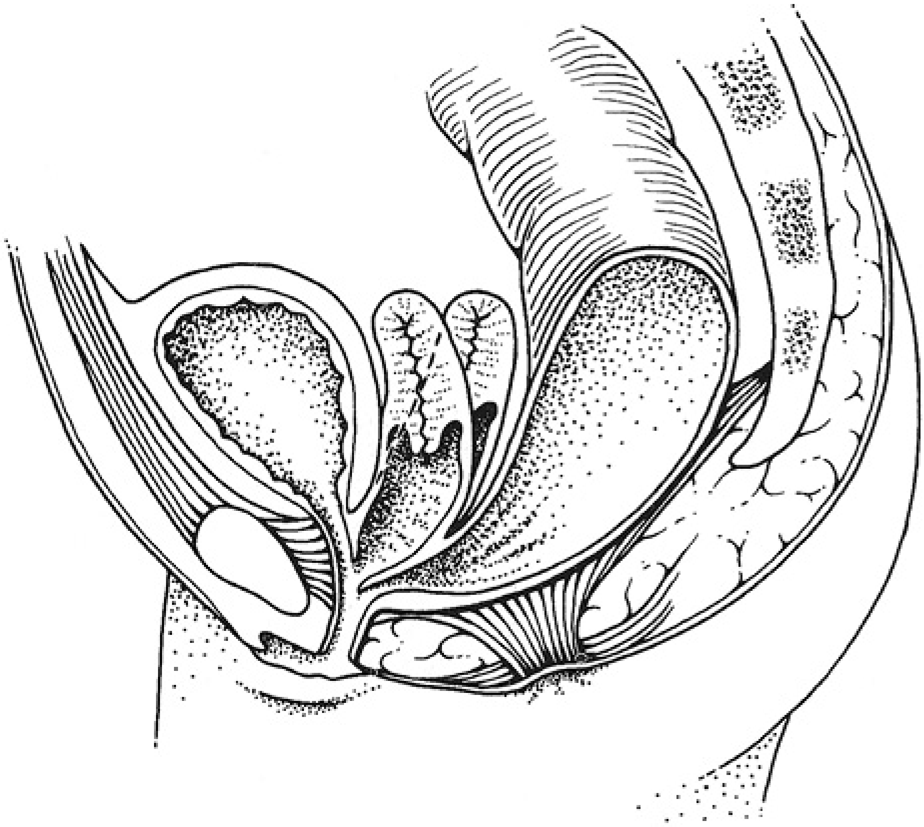


44.3

If a hydrocolpos is correctly identified and drained, obstruction of the urinary tract is usually relieved, making urinary diversions such as a vesicostomy, ureterostomy, or nephrostomy unnecessary. Rarely, a near-atresia of the urethra exists and a vesicostomy may be justified.

Attempts to drain the urinary tract through the single perineal orifice by way of intermittent catheterization or dilatations are not recommended, as it is unpredictable whether the catheter will enter the bladder or the vagina. This particularly applies in cases of the long common channel. Blind dilatations of the single external orifice may also provoke damage that can interfere with the future repair.

The vagina and uterus commonly show varying degrees of septation. The rectum usually opens between the two hemivaginas (**Figure 44.4**).



44.4

ASSOCIATED DEFECTS

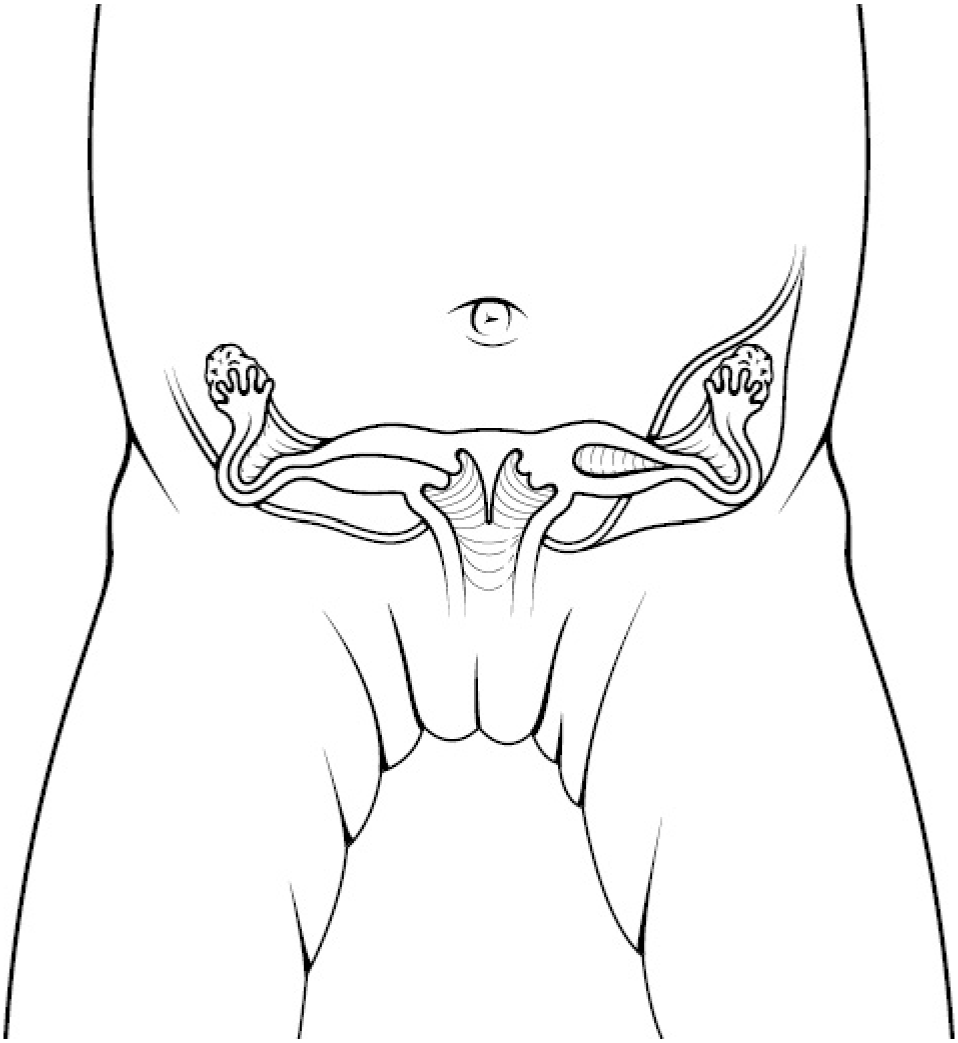
Genitourinary defects

Over 90% of cases have an associated genitourinary abnormality. Hydronephrosis, urosepsis, and metabolic acidosis represent the main source of morbidity and mortality in newborns with anorectal malformations. Thus, a thorough urologic investigation is mandatory.

Müllerian anomalies

About 60% of cases have duplication of Müllerian structures (double vagina and or uterus). This may have important gynecologic and obstetric implications and should be detected early in life, even if only to make the parents aware of their existence.

Cervical or vaginal atresias or stenoses may occur (**Figure 44.5**). When undetected, these may interfere with the drainage of menstrual blood during puberty causing hematometra, hematocolpos, or intra-abdominal pseudocysts from retrograde menstruation. The gynecologic anatomy can be ascertained during the main repair (if the abdomen is entered) or at the time of the colostomy closure.



44.5

OPERATIONS

Endoscopy

Thorough endoscopic examination (including vaginoscopy and cystoscopy) is essential for infants with cloaca to try to determine the anatomy. This can be done during the same anesthesia given to perform the initial colostomy and/or drainage of a hydrocolpos. The specific purpose of this procedure is to determine the length of the common channel, the status of the bladder neck, and the presence or absence of one or two vaginas and cervices.

Each type of cloaca has specific anatomic variants which represent a different technical challenge and must be recognized preoperatively.

Colostomy

All infants with a cloaca need a colostomy. It is important to perform the colostomy proximally enough to avoid it interfering with the repair of the malformation. In other words, the surgeon must leave enough redundant distal rectosigmoid to allow a pull-through, and for potential use of colon for vaginal replacement.

Drainage of the hydrocolpos

If present, the hydrocolpos needs drainage at the same time as the colostomy. If the hydrocolpos is not large enough to reach the abdominal wall above the bladder, it can be drained with a transabdominal indwelling catheter that should be left in place until the time of the main repair. A pigtail tube is recommended to avoid displacement of the catheter after the vagina is decompressed, becomes less inflamed, and recedes away from the abdominal wall. As a significant number of cases have two hemivaginas, the surgeon must be certain that the tube inserted into the hydrocolpos is really draining both of them. Occasionally, a window in the vaginal septum needs to be created in order to drain both with a single tube. Sometimes, the hydrocolpos is so large that it may even produce respiratory distress; such giant vaginas may be drained directly, connecting the vaginal wall to the abdominal wall as a tubeless vaginostomy.

Rarely, infants are unable to empty their bladders because they suffer from a near-atresia of the common channel. In such circumstances, they may require a vesicostomy or a suprapubic cystostomy.

DISTAL COLOSTOGRAPHY AND CLOACAGRAM

A high-pressure distal colostogram and injection of contrast through the single perineal orifice will help define the cloacal anatomy following postoperative recovery. This is ideally performed with contrast injected into urinary, gynecologic, and colorectal tracts, and can be viewed in three dimensions with a rotating fluoroscopy machine. This study can demonstrate the location of the rectum, and show the vagina or hemivaginas, and often assess for vesicoureteral reflux.

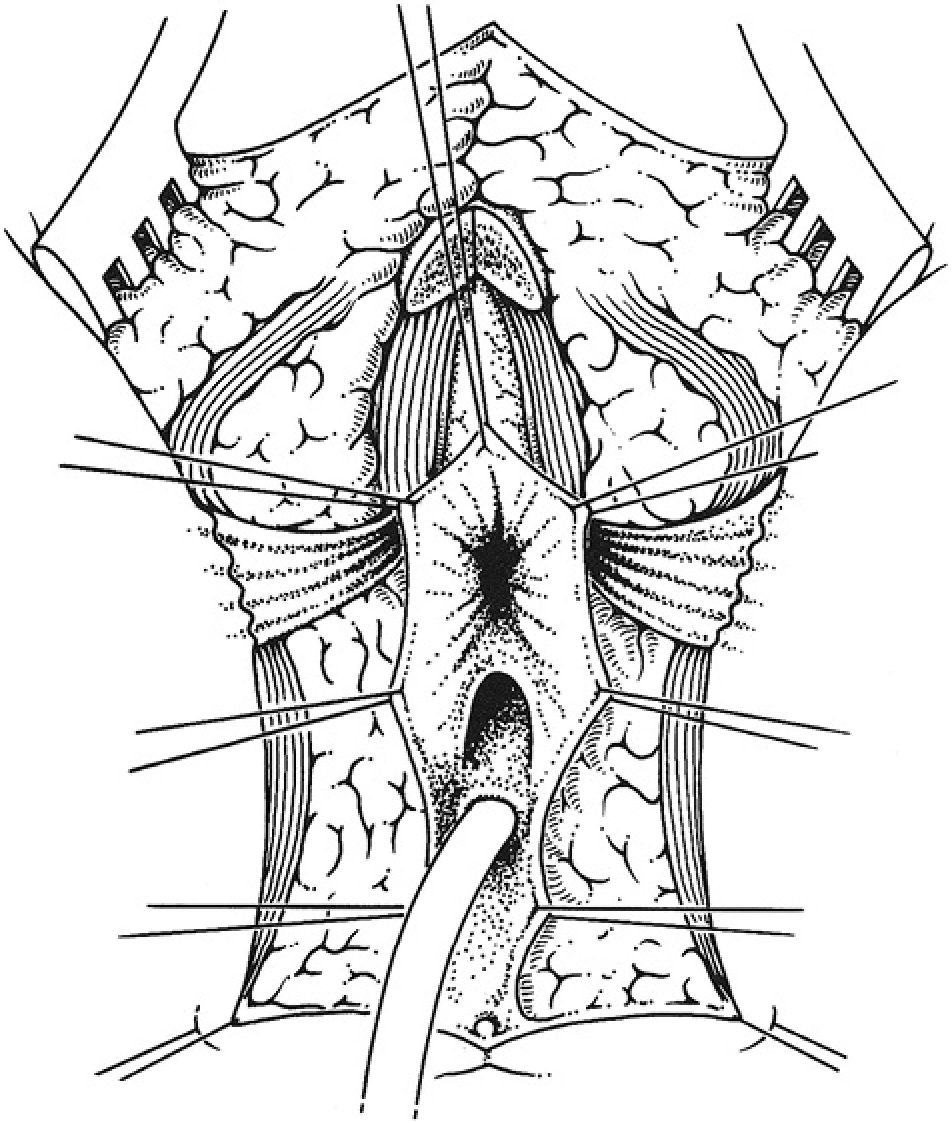
Definitive repair

The goal of the operation is to separate the rectum from the vagina and place it within the sphincteric mechanism. The urethra and vagina need to be mobilized so that each is a separate orifice at the perineum. The operation can be done once we have demonstrated weight gain after the colostomy opening.

CLOACAS WITH COMMON CHANNEL SHORTER THAN 3 CM

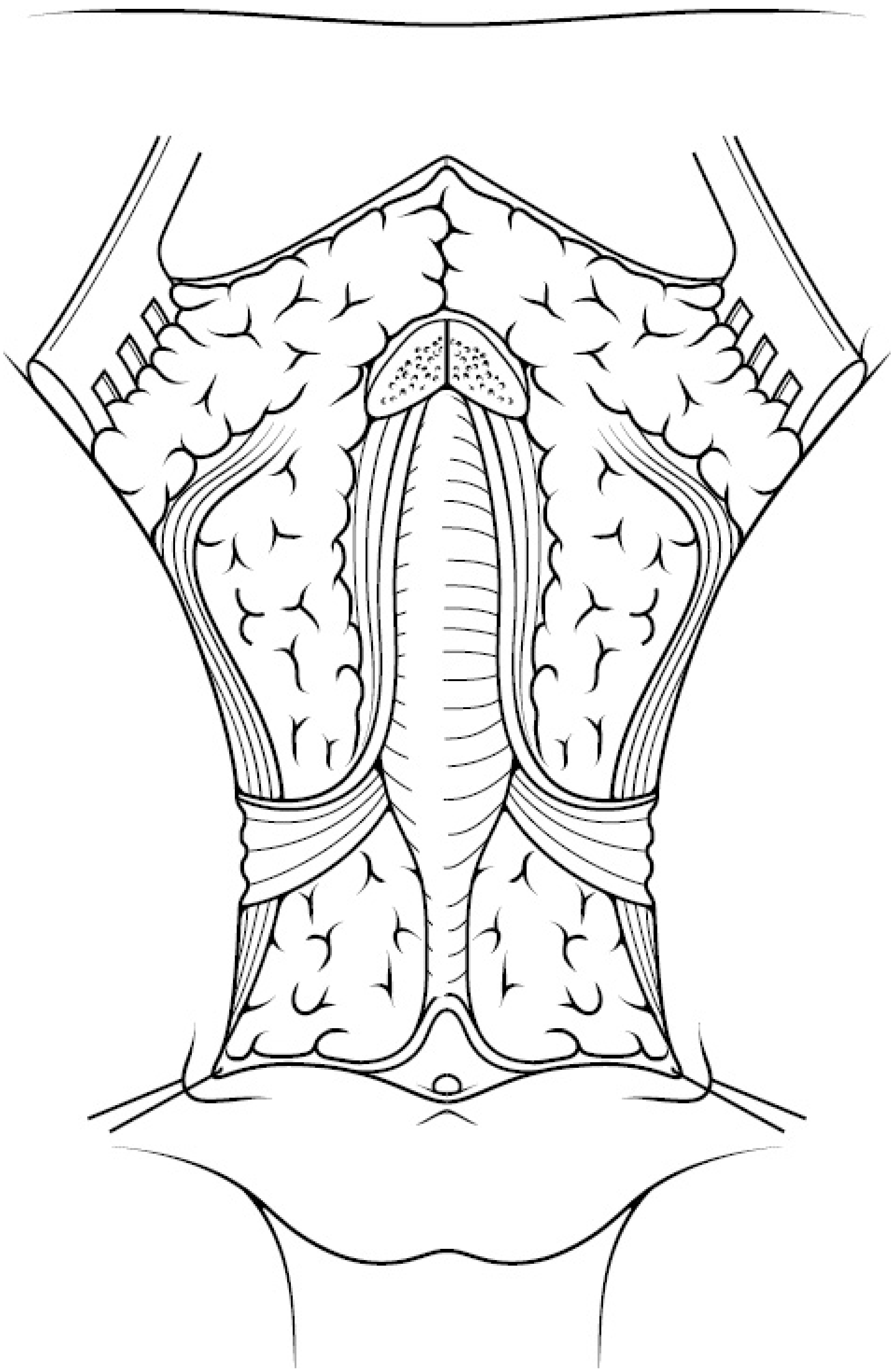
With the patient in the prone position, a long midsagittal incision is performed between the buttocks that extends from the middle portion of the sacrum through the sphincter mechanism and down into the single perineal opening. All of the muscle structures are divided in the midline.

The incision is continued all the way down to the single perineal orifice, exposing the entire malformation (**Figure 44.6**). The entire sphincter mechanism is divided in the midline.



44.6

The first visceral structure to be found is usually the rectum (**Figure 44.7**). The surgeon must be prepared to find bizarre anatomic arrangements of rectum, vagina, and urethra.

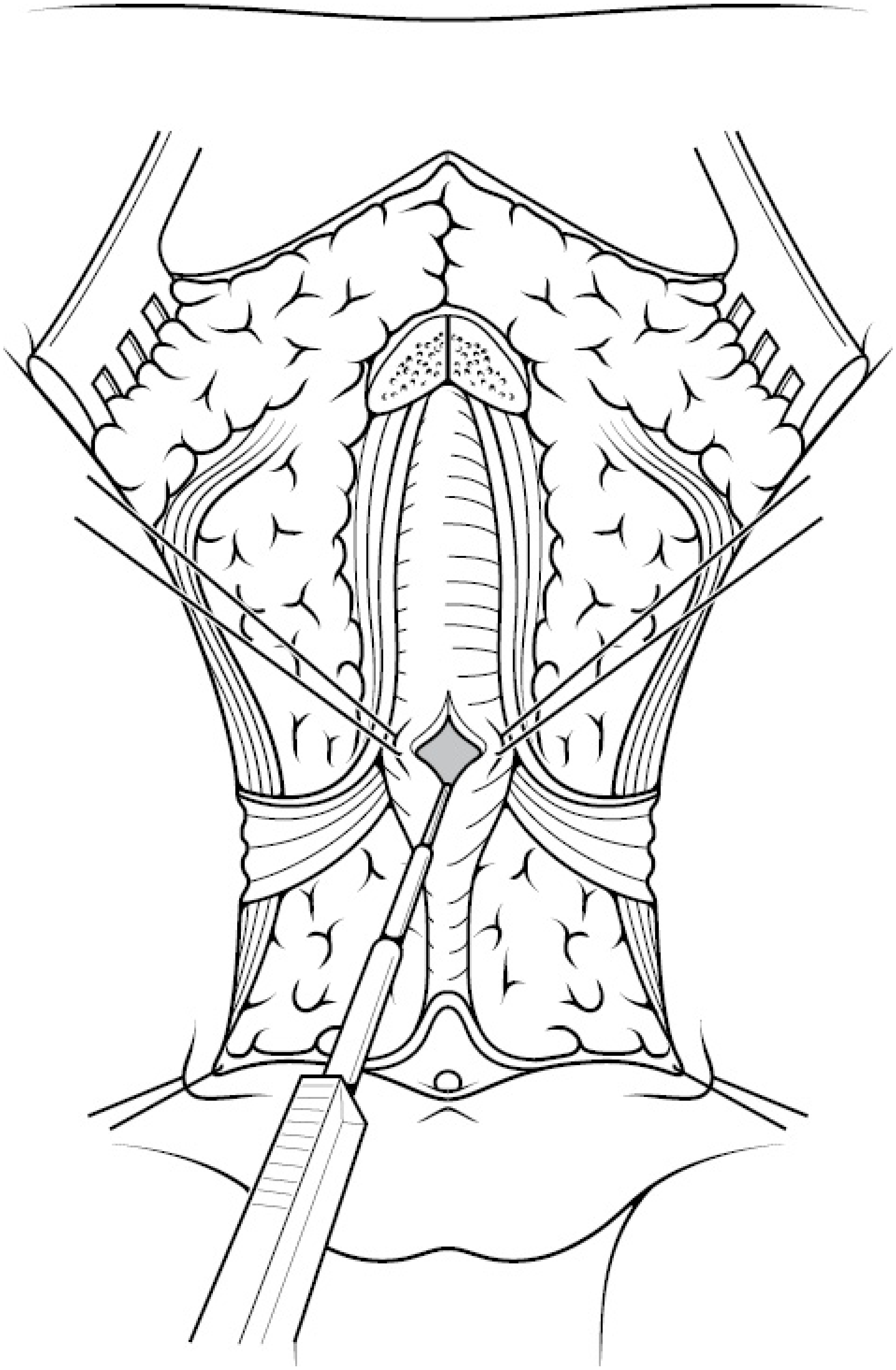


44.7

At this stage, the surgeon has an objective idea of the complexity of the defect and can directly measure the length of the common channel. If it is

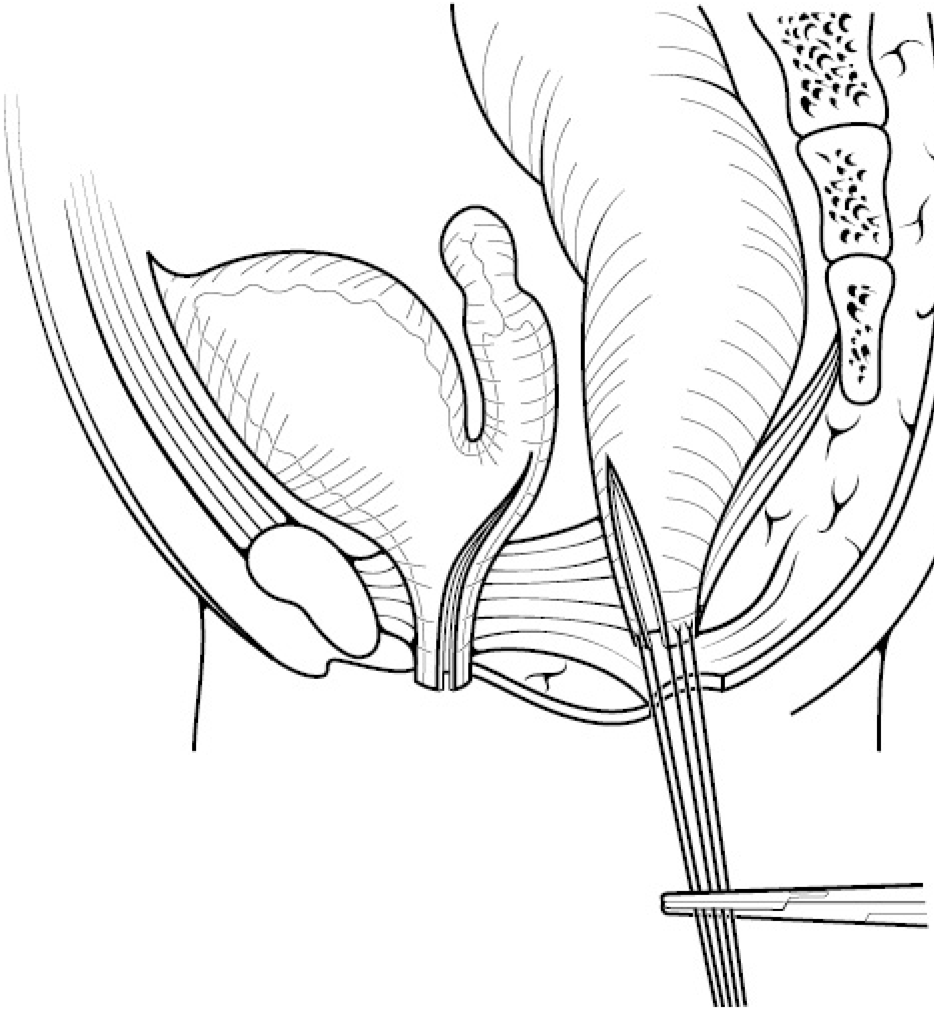
<3 cm, it will usually be possible to mobilize the entire urogenital sinus (vagina and urethra together, i.e. total urogenital mobilization), as well as the rectum, without opening the abdomen.

The rectum is opened in the midline and silk stay sutures are placed along the edges of its posterior, anterior and lateral walls (**Figure 44.8**).



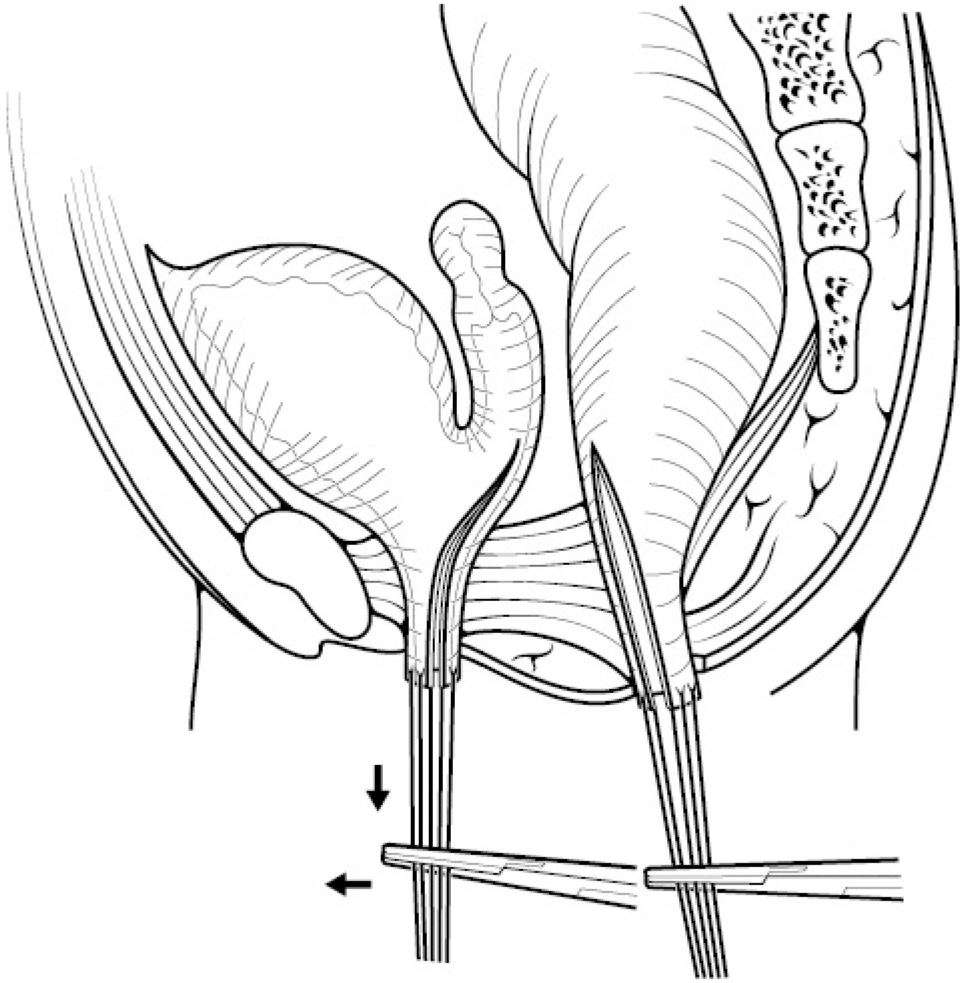
44.8

The next step consists of separating the rectum from the vagina, which may not be easy given they share a common wall. It is therefore necessary to make two walls out of one, working in a structure without a surgical plane for separation. To do this, it is necessary to apply uniform traction on the rectum, using the multiple sutures previously placed (**Figure 44.9**). We like to say that "traction creates the plane." In dealing with anorectal malformations, the surgeon must remember that about 70% of the operative time is spent in the separation of structures. This step is also the most technically demanding and requires a meticulous and delicate technique.



44.9

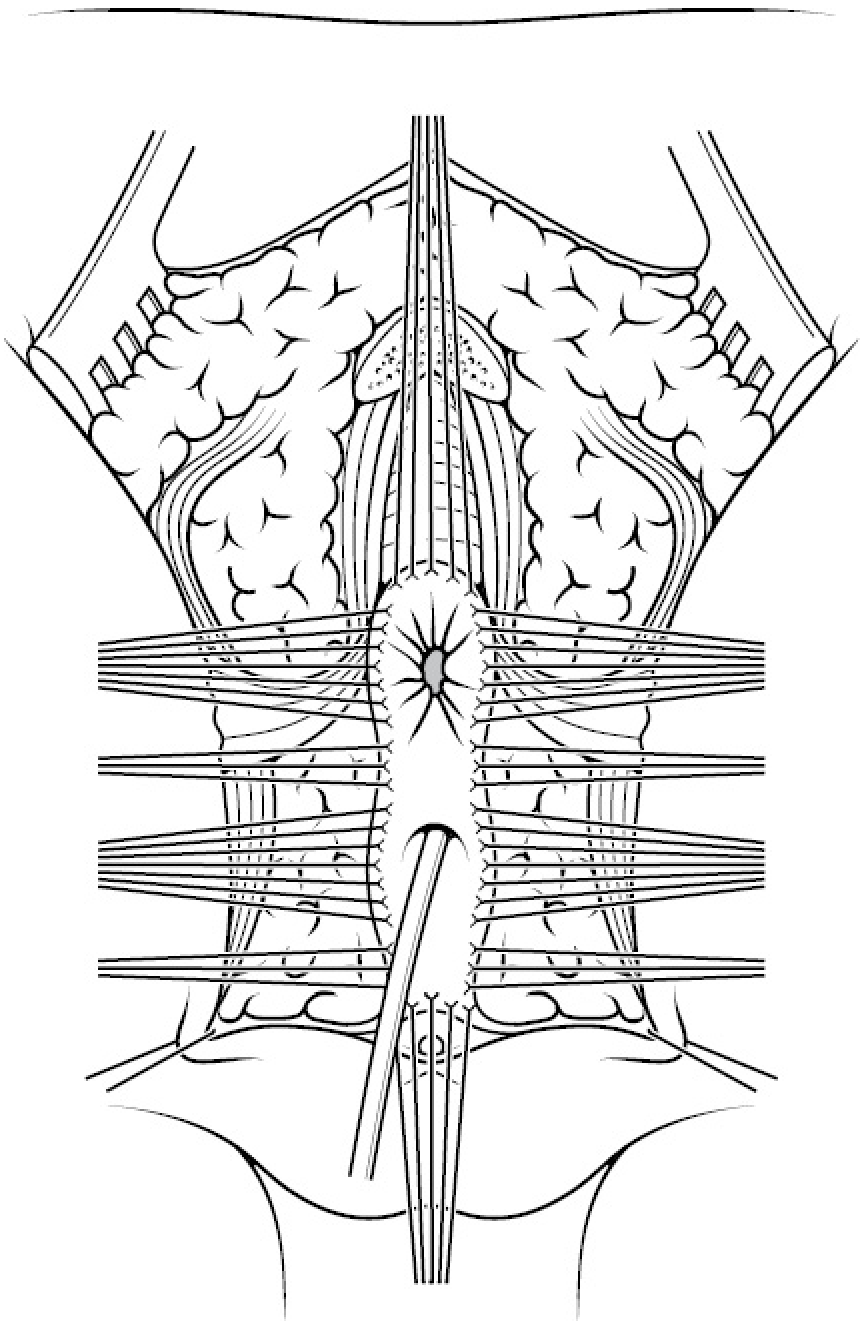
Once the rectum has been completely separated from the vagina, a circumferential, perirectal dissection is performed applying uniform traction, dividing bands and vessels that keep the rectum up in the pelvis (**Figure 44.10**). Special attention must be observed to avoid damaging or burning the rectal wall, since that would affect the intramural blood supply, provoking distal rectal ischemia. The dissection must continue until enough rectal length has been reached to be sure that the rectum reaches the perineum without tension.



44.10

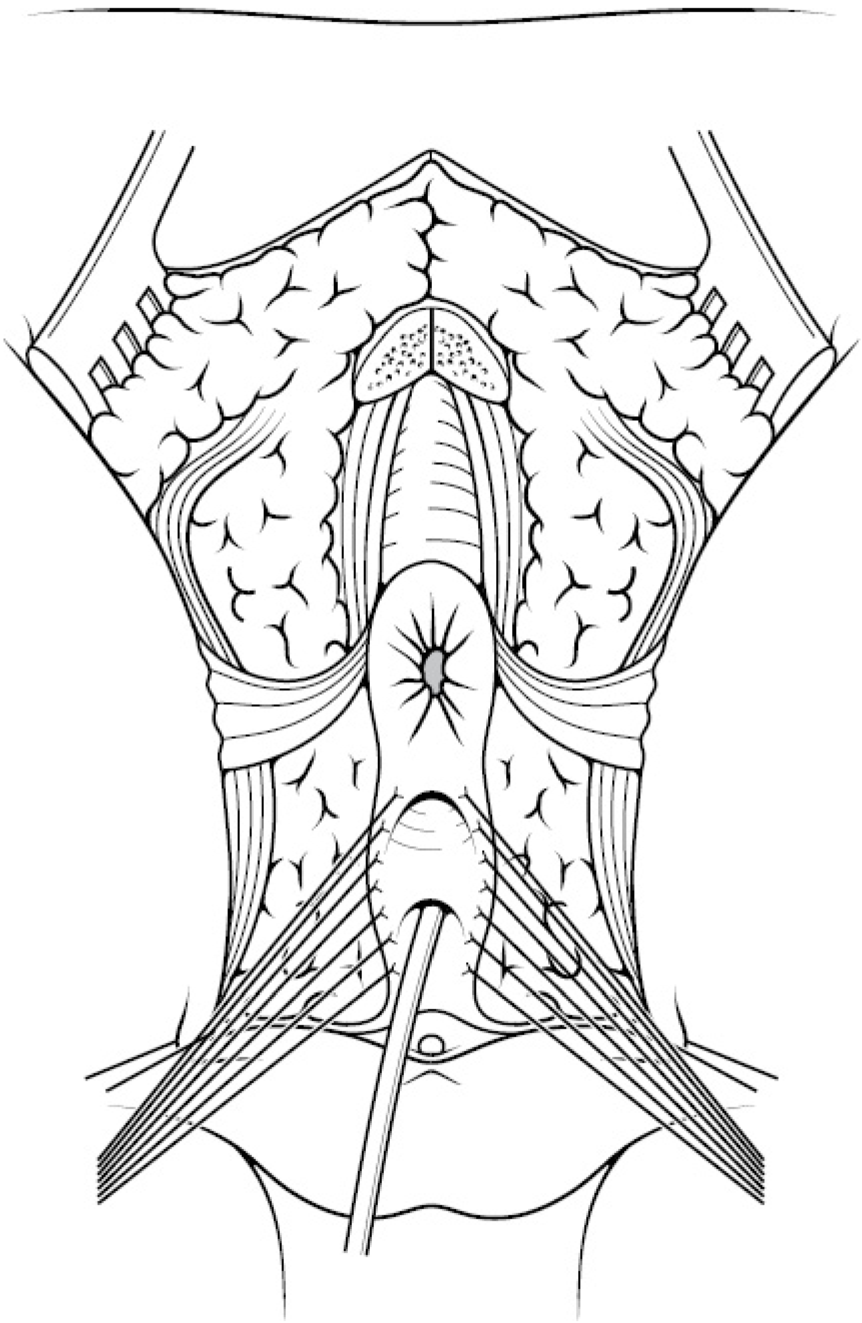
The next step of the operation consists in mobilizing together both urethra and vagina (total urogenital mobilization). For this, multiple silk stitches are placed on the edges of the vagina(s) and the common channel, in order to apply uniform traction on the urogenital sinus.

Another series of fine silk stay sutures is placed across the urogenital sinus about 5 mm proximal to the clitoris (**Figure 44.11**). This is transected full thickness between the last row of sutures and the clitoris. Taking advantage of the fact that there is a natural plane between the urogenital sinus and the posterior aspect of the pubis, a dissection is performed in between both structures. This dissection is bloodless; one can very rapidly reach the upper edge of the pubis. A fine grayish avascular membrane, which we call the "suspensory ligament of urethra and vagina," can be identified and divided, paying special attention to avoid touching important retropubic veins, which may produce significant bleeding. This maneuver immediately provides significant mobilization of the urogenital sinus and one can gain 2–3 cm of length.



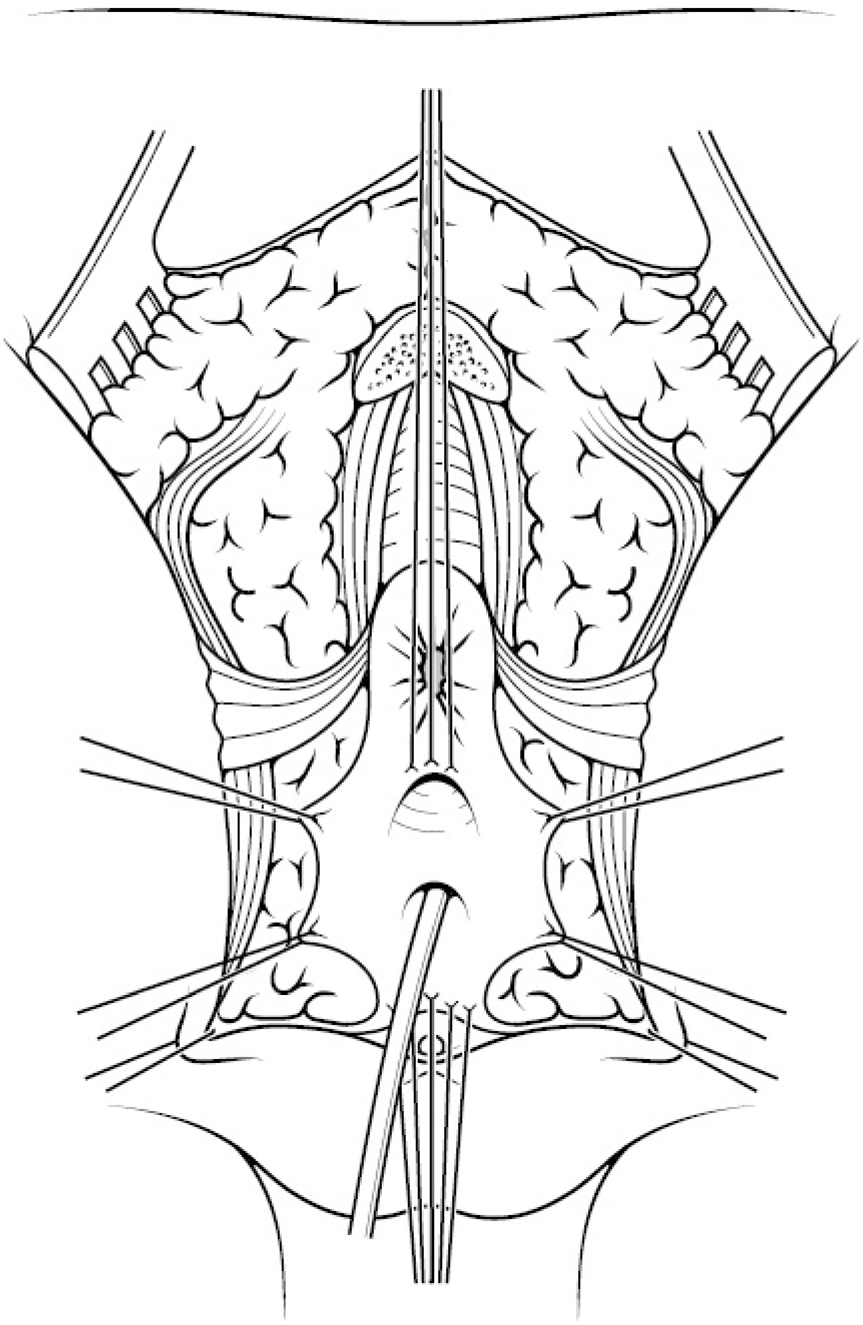
44.11

In addition, one can then dissect the lateral and dorsal walls of the vagina (**Figure 44.12**). This dissection is enough to repair about 60% of all cloacas.



44.12

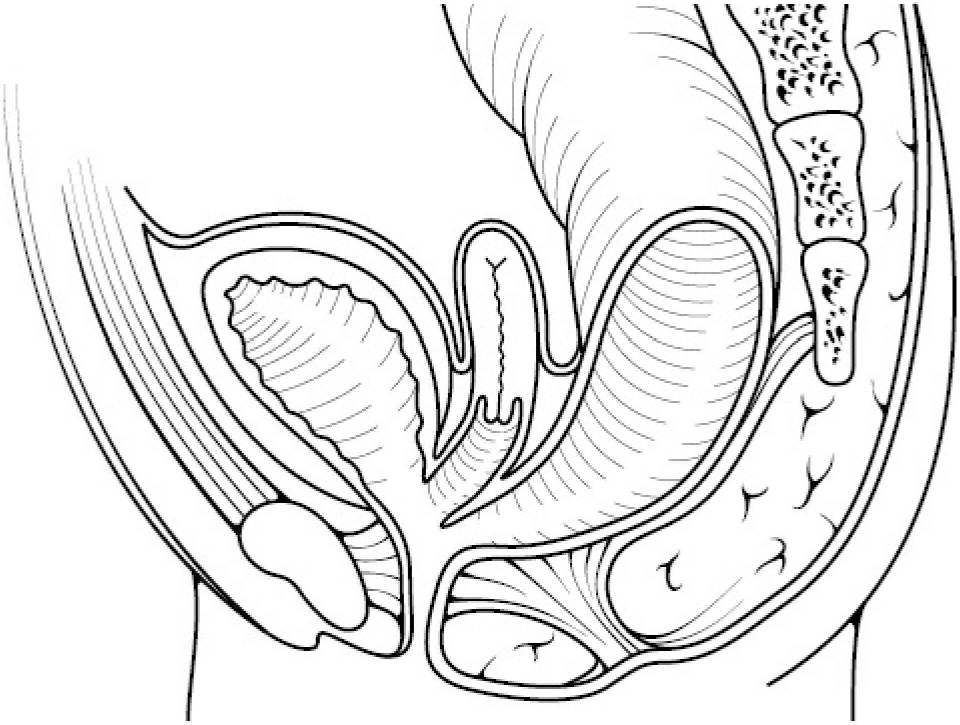
The urogenital mobilization has the additional advantages of preserving an excellent blood supply to both the urethra and vagina and of placing the urethral opening in a visible location to facilitate intermittent catheterization if necessary. This may be required in about 20% of cases. What used to be the common channel is divided in the midline, creating two lateral flaps that are sutured to the skin of the patient's new labia (**Figure 44.13**). The vaginal edges are mobilized to reach the skin to create the introitus. The limits of the sphincter are electrically determined. The perineal body is reconstructed, bringing together the anterior limit of the sphincter. The rectum is then placed within the limits of the sphincter and the anoplasty is performed.



44.13

CLOACAS WITH A COMMON CHANNEL LONGER THAN 3 CM

When endoscopy shows that the patient has a long common channel, the surgeon must be prepared to face a more significant technical challenge (**Figure 44.14**).



44.14

The patient is prepared with a total body preparation so that the surgeon can switch between the prone position and an abdominal approach, as described for recto-bladder neck fistula.

The operation starts with the posterior sagittal approach, and the rectum is separated from the vagina and urethra as described previously. The next step is to try to mobilize the urethra and vagina using the total mobilization maneuver, as also described previously. Occasionally, we are happily surprised to find that it is possible to perform the entire reconstruction in cases with a common channel of 3–4 cm. In most cases, however, a laparotomy is needed to complete the repair.

Occasionally, we find an extremely long common channel (≥5 cm). In such cases, the common channel should be left in place so that it can be used later for intermittent catheterization. In this situation, an attempt should be made to separate the vagina from the urinary tract, first from below and then from the abdomen. To do that, multiple 6/0 silk stay sutures are placed, taking the vaginal wall to apply uniform traction in an attempt to create a plane of dissection between the vagina and the urinary tract. This is a very delicate, meticulous (and tedious!) maneuver. With this dissection from the perineum, one can gain a separation of the vagina from the urinary tract for about 2 cm. The rest of the separation must be completed through the abdomen.

The patient is turned into a supine position. A midline laparotomy is recommended. The bladder is opened in the midline and feeding tubes are placed into the ureters to protect them. A long common wall between the vagina and the bladder exists, and both the ureters run through this common wall. The ureters sometimes must be skeletonized during this separation and therefore require protection.

The surgeon must be familiar with the different techniques of ureteral reimplantation because these patients may require this operation during the procedure. Once in the abdomen, the patency of the Müllerian structures can be confirmed by passing a 3 Fr feeding tube through the fimbriae of the Fallopian tubes and injecting saline solution. If one of the systems is not patent, we recommend its excision, with great care paid to avoiding damage to the blood supply of the ovary. When both Müllerian structures are atretic, we recommend leaving them in place and following the patient closely so that a decision can be made about them when she reaches puberty.

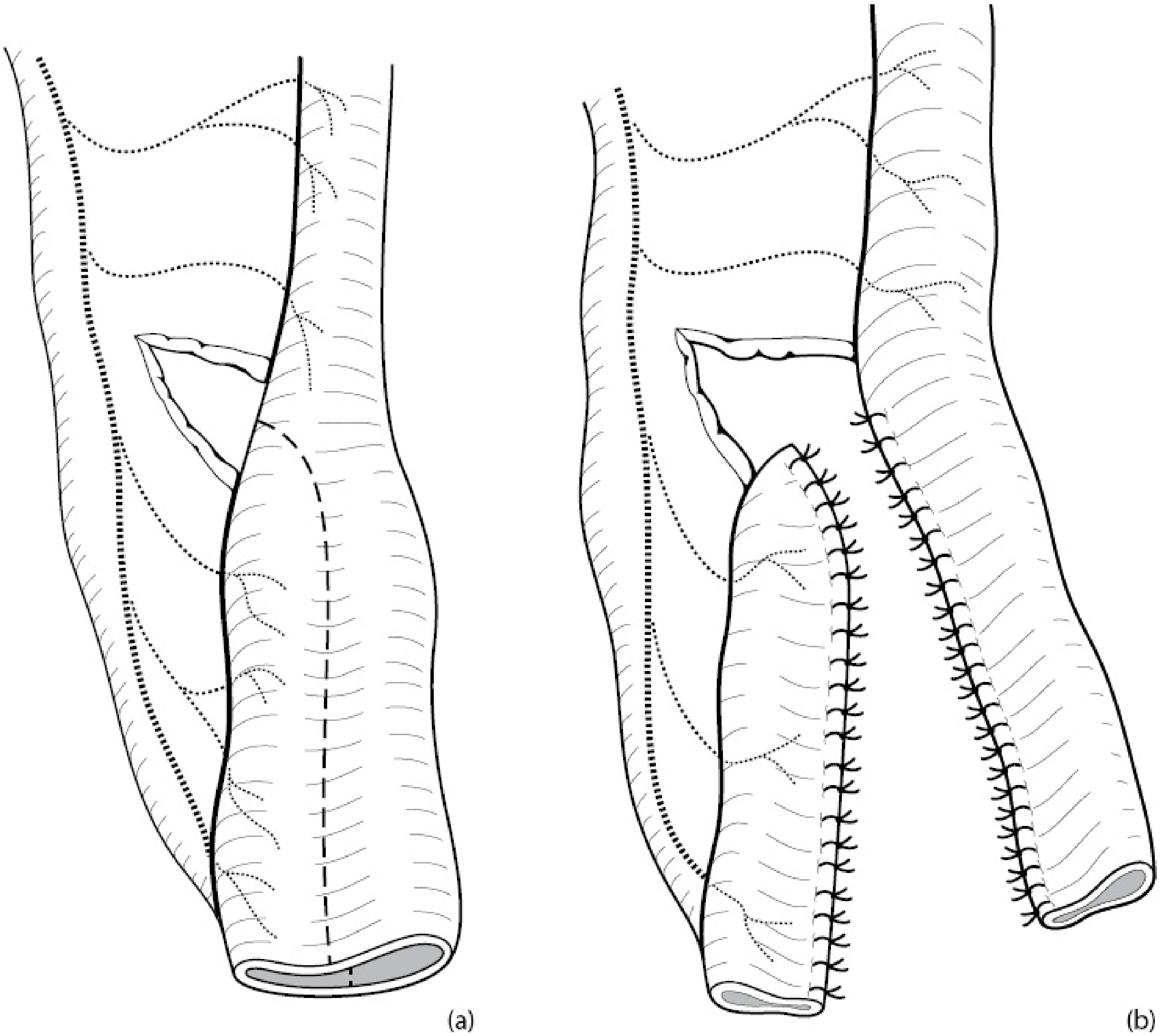
The procedure continues with the placement of traction sutures in the single uterus or in both hemiuteri. Traction sutures are also placed in the dome of the bladder. With the use of traction on both structures, dissection is initiated between the urinary tract and the vagina. This dissection is continued all the way down to meet the previous dissection initiated from below. The vagina is thus separated from the urinary tract, with care being taken to preserve its blood supply from the uterine vessels. Once the vagina(s) has/have been separated from the bladder and urethra, the surgeon should make decisions based on the specific anatomic findings. The vagina at this point may reach the perineum. If it is found to be too short, some form of vaginal replacement or vaginoplasty is required.

Vaginal replacement

The vagina can be partially or totally replaced with bowel when it is very small and is located very high, or in cases of absent vagina. The choices are rectum, colon, or small bowel.

RECTUM

Replacing the vagina with rectum is only feasible in patients who have no potential for bowel control or for those who have a megarectum that is large enough or long enough to be divided into a portion with its own blood supply, which will form the new vagina, and a portion with enough circumference to reconstruct an adequate-sized rectum. The blood supply of the rectum will be provided transmurally from branches of the inferior mesenteric vessels (**Figure 44.15a and b**).



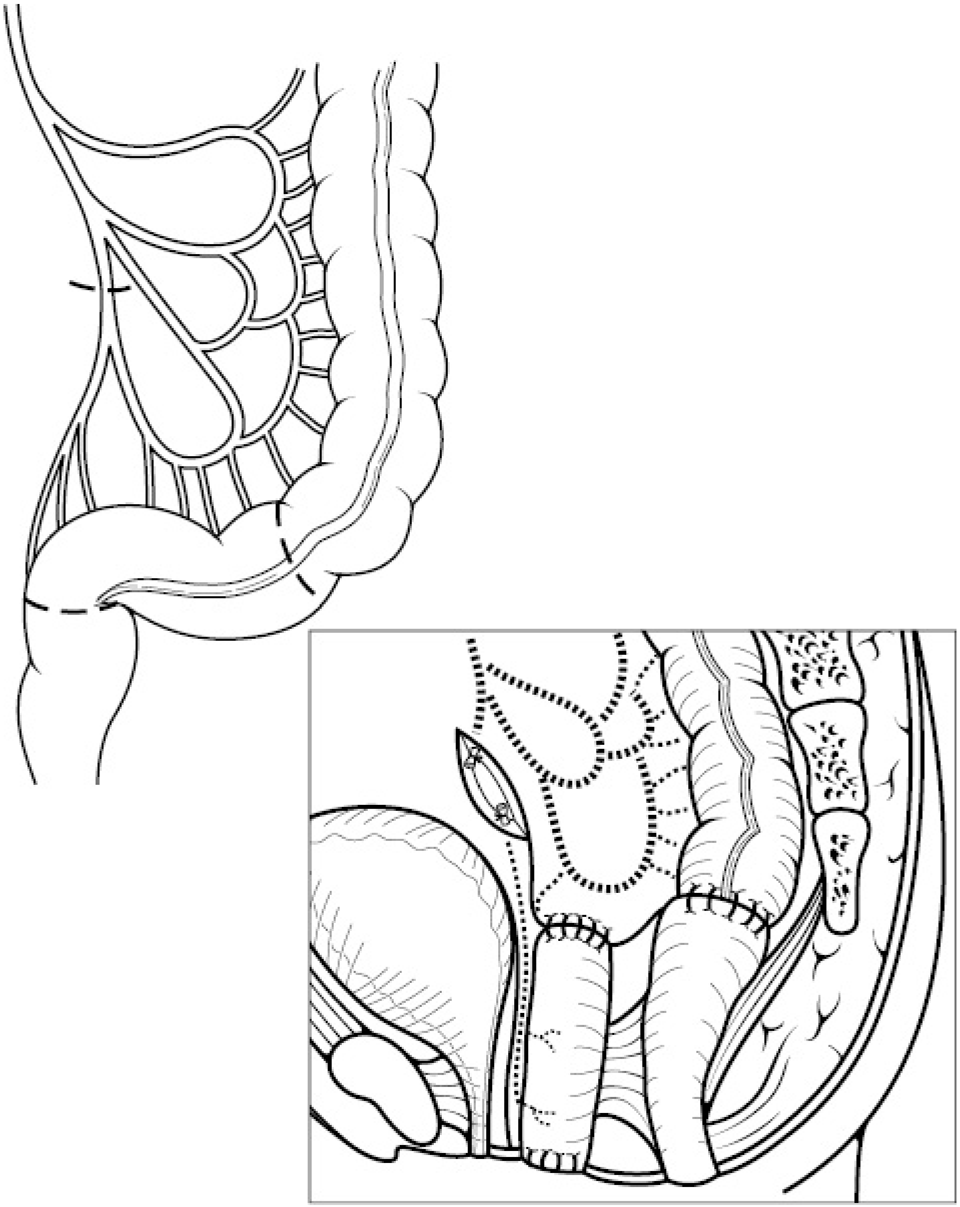
44.15a,b

COLON

In our opinion, the descending colon is the best part of the intestine to use to replace the vagina. The reasons are:

- It has excellent vascular arcades, which allow a relatively easy mobilization down to the perineum, without tension and preserving an excellent blood supply.
- It has a thick wall, which allows sexual intercourse without damage of the bowel wall.

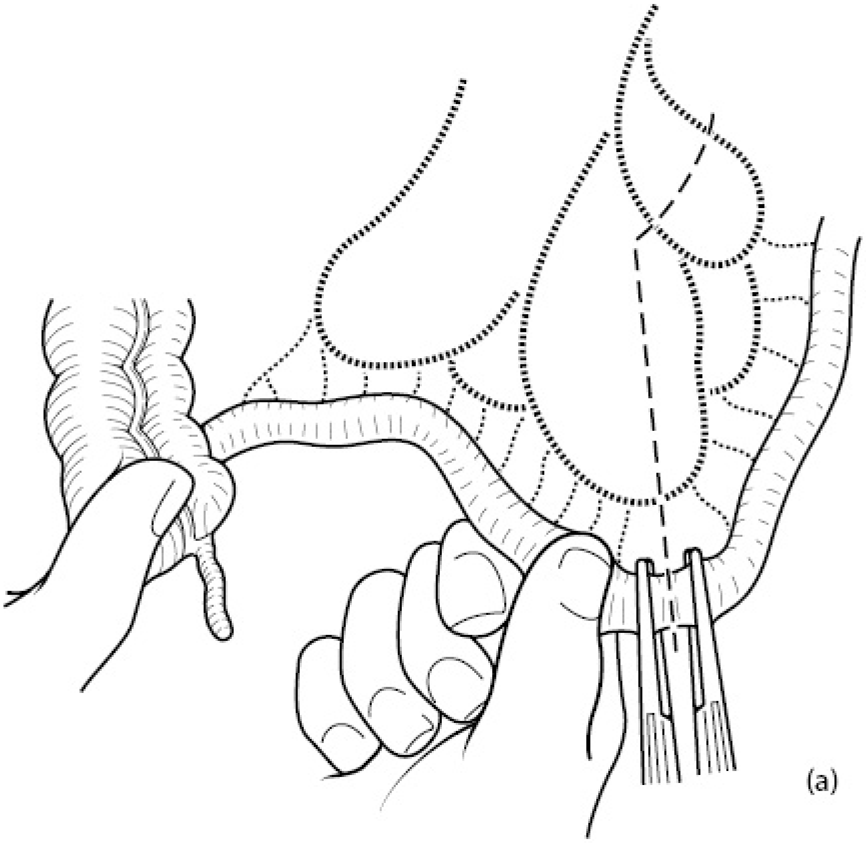
We specifically recommend a full exposure of the entire colon and its mesentery, to make an adequate decision as to which branches of the mesenteric vessels to ligate, preserving the vascular arcade to guarantee an excellent blood supply to the selected piece of colon. To achieve this, it is frequently necessary to mobilize both splenic and hepatic flexure of the colon. In cases of partial vaginal replacement, the selected piece of colon must be sutured to the native vagina and the patient may preserve a potential reproductive capacity. In cases requiring total vaginal replacement, the upper part of the selected piece of colon must be closed; the replacement will serve the function of having sexual activity but not reproductive purpose. The sigmoid can also be used as a colonic vaginal replacement (**Figure 44.16**). Sometimes, the colostomy interferes with the mobilization of specific portions of the colon and the surgeon must be creative enough to solve the challenge.



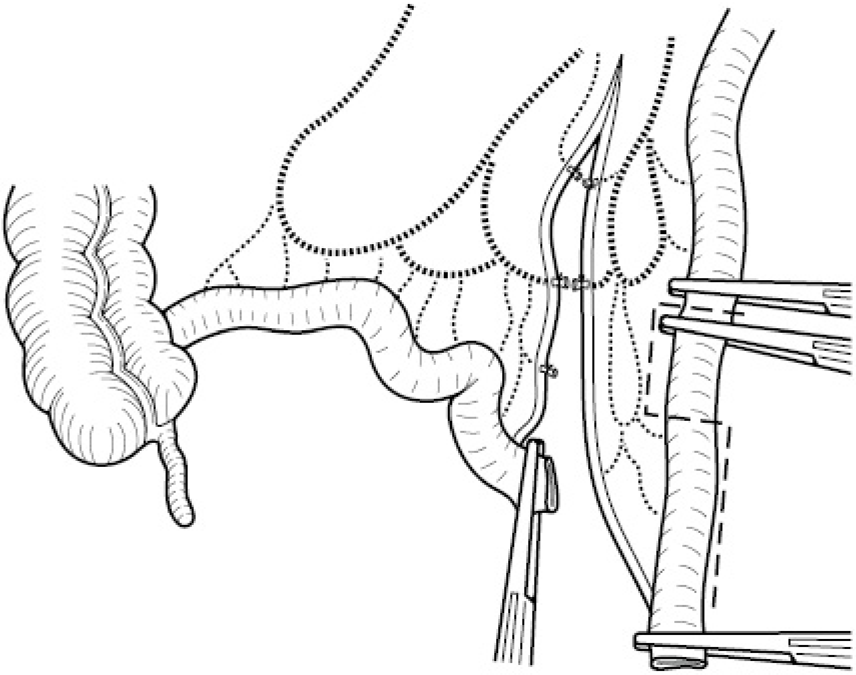
44.16

SMALL BOWEL

Small bowel is the least desirable option to replace the vagina. This is because its blood supply is very delicate, it may twist very easily, which will result in losing the piece of bowel chosen for reconstruction, and it has a very thin wall, which may be too delicate for sexual function. The mesentery of the small bowel is longer in an area approximately 15 cm proximal to the ileocecal valve (**Figure 44.17a**). A segment of this portion of the small intestine is selected, preserving its mesentery (**Figure 44.17b**).



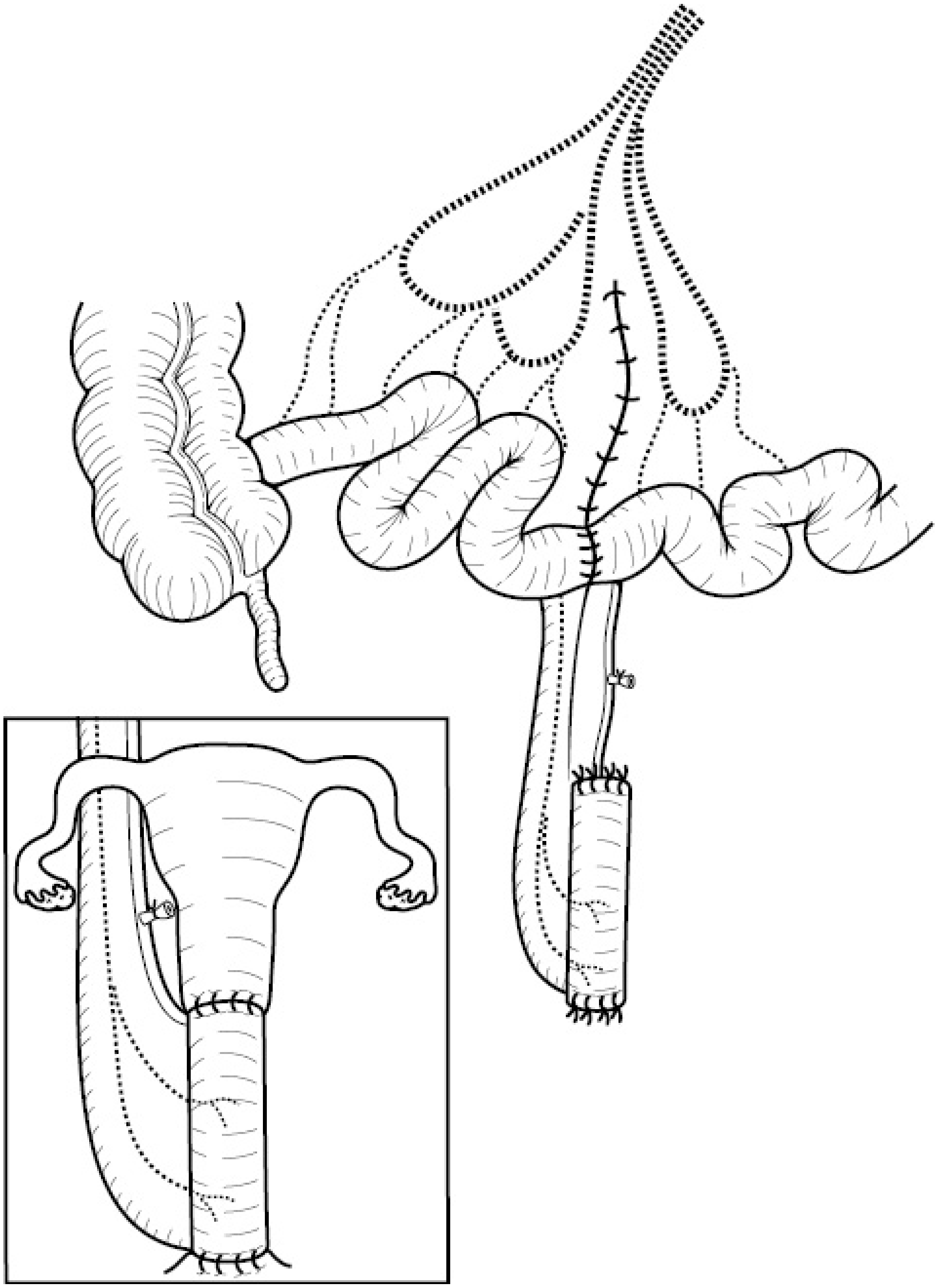
(a)



(b)

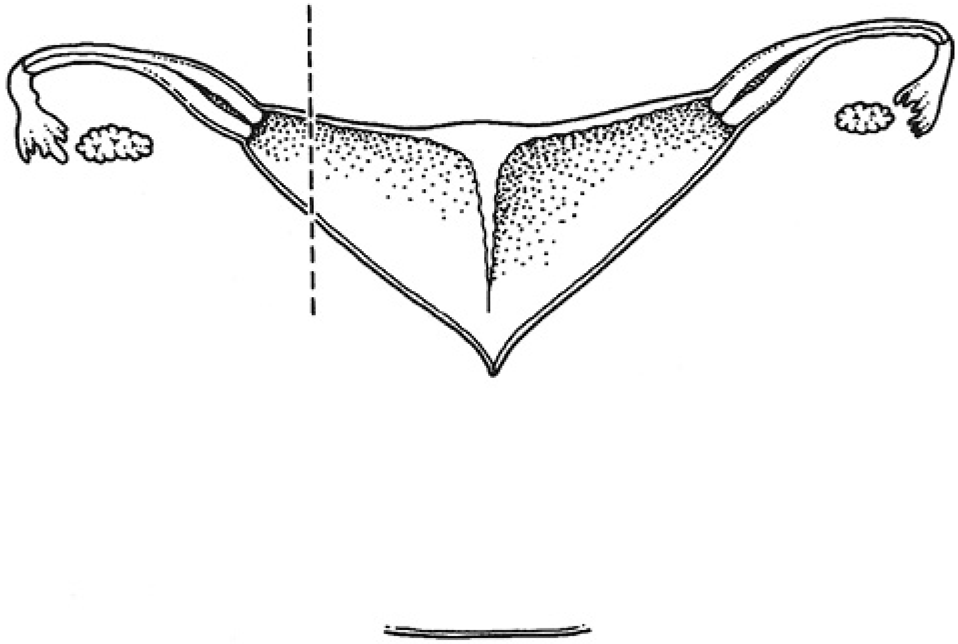
44.17a,b

The continuity of the small intestine is re-established with an end-to-end anastomosis. Two more anastomoses are necessary: The upper one between the segment of small intestine and the upper vagina, and the lower one between the lower part of the intestine and the perineal skin (new labia) (**Figure 44.18**).



44.18

During separation of the vagina from the urinary tract, the blood supply to both hemiuteri and hemivaginas reaches the hemiuterus laterally and must be preserved until a decision is reached concerning the type of vaginal mobilization to be performed (**Figure 44.19**).

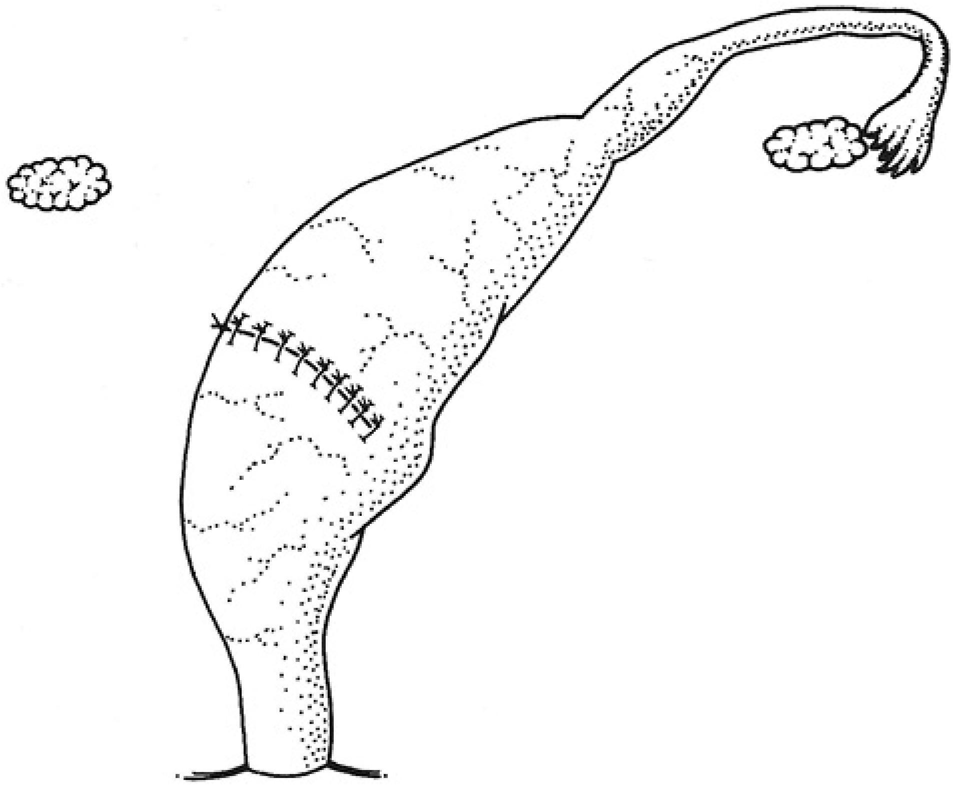


44.19

After separation of the vagina from the urinary tract has been completed, the surgeon may find out that the vagina is located too high in the pelvis to be mobilized down to the perineum. In such a case, the patient will require a vaginal replacement or, in specific cases, the surgeon may decide to use a technical maneuver called “vaginal switch.”

The vaginal switch maneuver is applicable in cases with a specific anatomic variant consisting of a long common channel and two dilated hemivaginas as a consequence of bilateral hydrocolpos and two hemiuteri widely separated. The distance between one hemiuterus and the other may be long enough to reach the perineum, provided one of the hemiuteri is sacrificed. One of the hemiuteri and the ipsilateral Fallopian tube are resected, with particular care given to preserving the blood supply of the ovary. The blood supply of the hemivagina of that side is sacrificed; that of the contralateral hemivagina is preserved and is sufficient for both hemivaginas. The vaginal septum is resected, and both hemivaginas are tubularized into a single vagina, taking advantage of their combined long lateral dimension.

What used to be the dome of the hemivagina where the hemiuterus was resected is then turned down to the perineum (**Figure 44.20**). This is an excellent maneuver, but it can only be performed when the anatomic characteristics fulfill the requirements described.



44.20

COMPLICATIONS

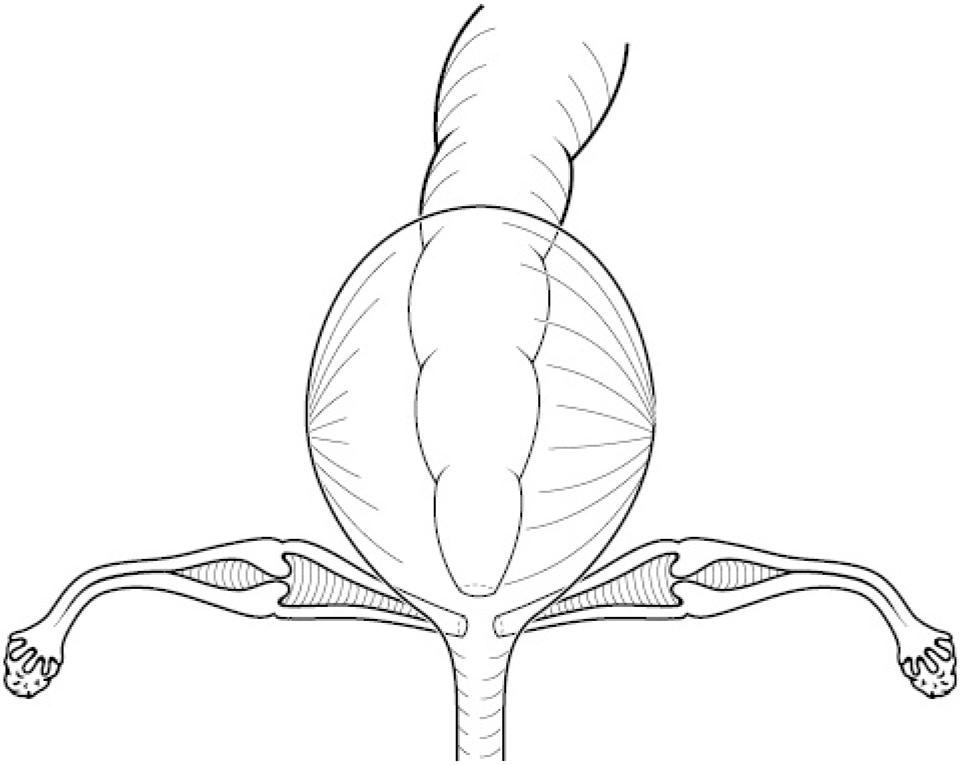
Urethrovaginal fistula

In the past, urethrovaginal fistula was the most common and feared complication in cases of persistent cloaca. In those years, the repair of cloacas included the separation of the vagina from the urethra; by definition, we left a partially damaged anterior vaginal wall, located in front of the neourethra suture line, which represents a high risk for a fistula formation. With the advent of the "total urogenital mobilization" maneuver, this complication has essentially been eliminated.

Acquired vaginal atresia

Ischemic vaginal fibrosis can occur secondary to an excessive dissection in an unsuccessful attempt to mobilize a very high vagina. To avoid this complication, adequate mobilization without tension or one of the described vaginal replacement maneuvers should be selected.

In the highest type of cloaca, one may find two little hemivaginas attached to the bladder neck or even to the trigone of the bladder (**Figure 44.21**). In these cases, the rectum also opens in the trigone. The separation of these structures is carried out through the abdomen. Unfortunately, when that separation is completed, patients are often left with no bladder neck or with a very severely damaged bladder neck. At that point, the surgeon must have enough experience to decide whether to reconstruct the bladder neck or to close it permanently. In the first situation, most patients will need intermittent catheterization to empty the bladder, and there is no guarantee that the bladder neck reconstruction will work. In the second situation (permanent closure of the bladder neck), a vesicostomy is created, and the patient will require a continent diversion-type of procedure at the age of urinary continence (bladder augmentation and Mitrofanoff type of conduit). In this particular type of malformation, the patient also needs a vaginal replacement, which should be done as described previously.



44.21

POSTOPERATIVE MANAGEMENT AND COLOSTOMY CLOSURE

Postoperatively, patients generally have a smooth course. Pain is not a problem, except for those who have undergone a laparotomy.

The Foley catheter is usually retained for 2–3 weeks. In our experience, about 20% of those with a common channel <3 cm will require intermittent catheterization to empty the bladder. This proportion rises to about 80% in those with common channels >3 cm. We therefore leave the catheter in place until all the genitalia edema has disappeared and the urethral orifice is visible. The catheter is removed in the clinic and then the patient is observed to see if she can empty the bladder. If not, we teach the parents to pass the catheter intermittently. In cases of very long common channels, we prefer to leave a vesicostomy or suprapubic tube in place at the time of the cloaca repair.

If the surgeon anticipates that the infant will need bladder drainage for more than 2 weeks and will be able to pass urine spontaneously, we use a

Foley catheter. If the estimate is that the patient will need this for between 2 weeks and 3 months and has no vesicoureteral reflux, we recommend a suprapubic catheter. In cases with complex cloacas, poor sacrum, presence of tethered cord, and vesico-ureteral reflux, it is convenient to open a vesicostomy, to protect the kidneys. Those patients will eventually need a full urologic evaluation, including a urodynamic study later in life, in order to plan the urinary reconstruction and a ureterovesical reimplantation.

Intravenous antibiotics are administered for 24–48 hours after the operation. Prophylactic antibiotics are administered orally to avoid urinary tract infections while the catheter is in. Antibiotic ointment is applied locally for 5 days. Most patients go home after 2 days, or after 3–4 days for those who required a laparotomy.

Two weeks after the repair, anal dilatations are started, following the protocol described for all our patients with anorectal malformations.

For patients who had a suprapubic tube, a cystostogram is performed 1 month following surgery to verify the patency of the urethra and rule out the possibility of urethrovaginal fistulas or a urethral stricture. Intermittent clamping of the tube is commenced, and the residual urine is measured as an indicator of the efficiency of bladder function. The suprapubic tube remains in place until we have evidence of a good bladder function or the caregiver learns to catheterize the bladder when required. A vesicostomy can be similarly used.

At the time of colostomy closure, an endoscopy should be performed to ensure that the repair is intact. If the cloacal repair did not require an abdominal approach, inspection of the Müllerian structures (as already described) should be performed at the time of colostomy closure.

OUTCOME

A patient who has one to three bowel movements per day, remains clean between bowel movements, and shows evidence of feeling or pushing during bowel movements, has a good bowel movement pattern and usually a good prognosis. This type of patient is trainable. A patient with multiple bowel movements or one who passes stools constantly without showing any signs of sensation or pushing usually has a poor functional prognosis.

Cloacas represent a spectrum of defects that can be subclassified on the basis of potential for bowel and urinary control, for which the length of the common channel seems to be an important prognostic factor, along with the status of the patient's sacrum and spine.

Many patients suffer from a deficient emptying mechanism of the bladder. However, they do not have the typical "Christmas-tree" type of neurogenic bladder of spina bifida patients, but rather have a flaccid, smooth, large bladder that does not empty completely. Fortunately, most patients with cloacas have a very good bladder neck. The combination of a good bladder neck and a floppy, flaccid bladder makes these patients ideal candidates for intermittent catheterization, which keeps them completely dry. There are two exceptions. One is patients who have a very long common channel, in which the hemivaginas are attached to the bladder neck, and, after these structures have been separated, the patients are left with no or a severely damaged bladder neck. The second exception is a small number of patients who are born with separated pubic bones. These patients do not have a bladder neck and they eventually require a continent diversion type of operation. These patients could be described as having a covered exstrophy.

Table 44.2 shows the clinical results obtained in our series of cases with cloacal malformations.

Table 44.2 Clinical results in cloacal surgery (based on our series of 539 cloacas, 228 of whom were available for follow-up and above the age of potty training)

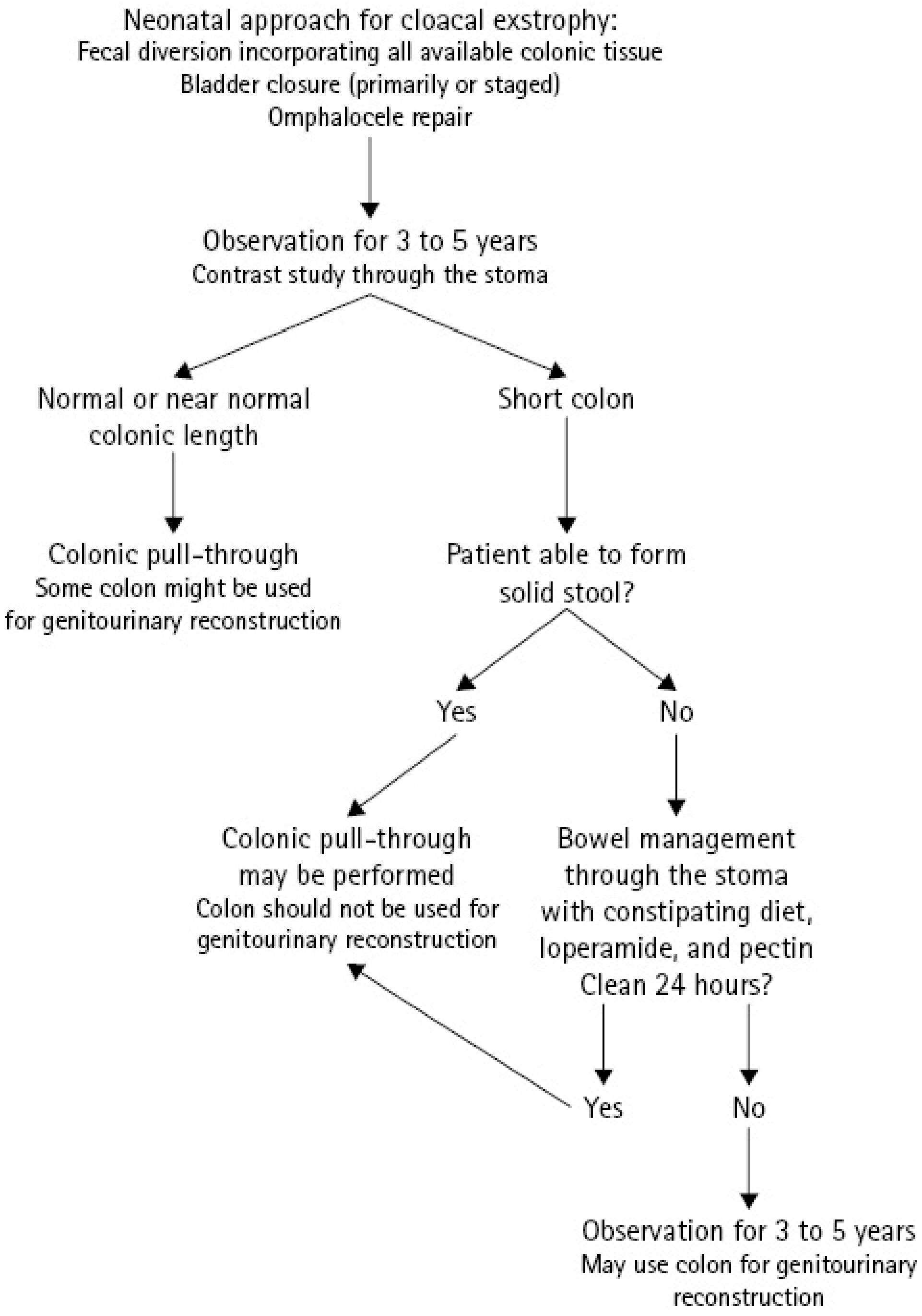
Common channel	Short, <3 cm (%)	Long, >3 cm (%)
Bowel function		
Total cases evaluated	**127**	**101**
Voluntary bowel movement	65/99 (66%)	26/73 (36%)
Totally continent[a]	34/65 (52%)	7/26 (27%)
Soiling	61/95 (64%)	47/56 (84%)
Constipated	38/98 (39%)	17/61 (28%)
Urinary function		
Normal	76/103 (74%)	28/87 (32%)
Dry with intermittent catheterization	23/103 (22%)	56/87 (64%)
Through the urethra	18/23 (78%)	28/56 (50%)
Through Mitrofanoff conduit	5/23 (22%)	28/56 (50%)

[a] Voluntary bowel movement and no soiling.

CLOACAL EXSTROPHY

Cloacal exstrophy represents the most complex of all anorectal and genitourinary malformations. The initial approach consists of the repair of the omphalocele, closure of the bladder (primarily or staged) with or without osteotomies, and the creation of an end colostomy utilizing all available colon. We then observe the patient for 3–5 years, since the ultimate goal for these patients should be to achieve dryness for urine, cleanliness for stool, and sexual function. With respect to fecal continence, most of these patients have a poor prognosis due to the presence of a dysplastic sacrum, significant vertebral defects, suboptimal anal sphincter and perineal muscle function, and different degrees of colon length, usually short, which leads to loose stool. Despite this poor prognosis, it is the capacity to form solid stool that allows for a successful bowel management program, where the child is kept clean for 24 hours with the use of one enema a day. To achieve this, it is vital that every piece of hindgut, no matter how small, is incorporated into the fecal stream, because it has been noted that these apparently useless small segments of colon will grow considerably with use and time. Only after determining the potential for bowel control should a urological reconstruction be planned.

With a unified approach that includes the participation of a team of dedicated surgeons (pediatric surgeon, pediatric urologist, pediatric orthopedic surgeon, and neurosurgeon), most patients born with cloacal exstrophy can have a pull-through and are kept clean with bowel management, and dry with intermittent catheterization, allowing for a better quality of life (**Figure 44.22**).


Neonatal approach for cloacal exstrophy:
Fecal diversion incorporating all available colonic tissue
Bladder closure (primarily or staged)
Omphalocele repair
Observation for 3 to 5 years
Contrast study through the stoma
Normal or near normal colonic length
Short colon
Colonic pull-through
Some colon might be used for genitourinary reconstruction
Patient able to form solid stool?
Yes
No
Colonic pull-through may be performed
Colon should not be used for genitourinary reconstruction
Bowel management through the stoma with constipating diet, loperamide, and pectin
Clean 24 hours?
Yes
No
Observation for 3 to 5 years
May use colon for genitourinary reconstruction

44.22 Decision-making algorithm for the management of newborns with cloacal exstrophy.

FURTHER READING

Bischoff A. The surgical treatment of cloaca. *Semin Pediatr Surg* 2016; 25: 102–7.

Bischoff A, Levitt MA, Breech L, et al. Hydrocolpos in cloacal malformations. *J Pediatr Surg* 2010; 45: 1241–5.

Bischoff A, Levitt MA, Foong YL, et al. Prenatal diagnosis of cloacal malformations. *Pediatr Surg Int* 2010; 26: 1071–5.

Breech L. Gynecological concerns in patients with anorectal malformations. *Semin Pediatr Surg* 2010; 19: 139–45.

Hendren WH. Further experience in reconstructive surgery for cloacal anomalies. *J Pediatr Surg* 1982; 17: 695–717.

Levitt MA, Bischoff A, Peña A. Pitfalls and challenges of cloaca repair: How to reduce the need for reoperations. *J Pediatr Surg* 2011; 46: 1250–5.

Levitt MA, Mak GZ, Falcone RA, Peña A. Cloacal exstrophy – pull through or permanent stoma? A review of 53 patients. *J Pediatr Surg* 2008; 43: 164–70.

Levitt MA, Peña A. Pitfalls in the management of newborn cloacas. *Pediatr Surg Int* 2005; 21: 264–9.

Peña A. Total urogenital mobilization – an easier way to repair cloacas. *J Pediatr Surg* 1997; 32: 263–8.

Soffer SZ, Rosen NG, Hong AR, et al. Cloacal exstrophy: A unified management plan. *J Pediatr Surg* 2000; 35: 932–7.

45

Laparoscopic surgery and imaging-directed surgery for anorectal malformation

MATTHEW W. RALLS AND MARCUS D. JARBOE

PRINCIPLES

The incidence, classification, and decision-making algorithms for the spectrum of anorectal malformations (ARMs) are detailed in Chapter 43. The authors will focus this chapter on the application of minimal access techniques to the repair of ARMs.

Indications

The laparoscopic approach is not indicated for children with low ARMs characterized by a perineal or vestibular fistula. In general, laparoscopic-assisted anorectal pull-through (LAARP) is most appropriate for high ARMs in boys, including those with rectourethral, prostatic, or bladder neck fistulas, and high imperforate anus without a fistula. Laparoscopy has also been used by several authors for selected forms of cloacal malformations, in order to evaluate the intra-abdominal anatomy and to perform mobilization of pelvic structures. The role of laparoscopy in this setting will not be addressed in this chapter.

The choice of posterior sagittal anorectoplasty (PSARP) versus LAARP remains controversial and somewhat surgeon-specific. Some advocate the laparoscopic approach for only the highest ARMs (proximal fistulas in the bladder neck or prostatic urethra) in which the PSARP is more difficult, and believe that outcomes are superior with this approach. In contrast, PSARP for lower bulbar urethral fistulas is relatively easy, and the laparoscopic approach is correspondingly more difficult. Thus, some would argue PSARP is a better option for those patients. Others, including the authors, use laparoscopy for nearly all cases of male ARM without an external anal opening. This allows for less dissection of the subcutaneous, superficial, and deep sphincters muscles as well as the levators. Given the relative rarity of the disease process as well as the operative variability among and even within centers, there are no long-term data to suggest the most appropriate indication and/or contraindication for use of laparoscopy in children with ARM. In children with high ARM without a fistula, the decision should be made based on the level of the rectal atresia and surgeon comfort with laparoscopy.

Though not universally applied, some centers use magnetic resonance image (MRI) guidance to ensure appropriate directional placement of the pull-through segment through the perineum and pelvic musculature. MRI has been used preoperatively and postoperatively in a diagnostic capacity in children with ARM for many years. With the increased acceptance of LAARP, complications such as poorly positioned anal openings as well as malpositioned neoanal canals (lateral to the muscle complex) were increasing. Using MRI guidance at the time of the pull-through allows an understanding of the individual's luminal and muscular pelvic anatomy. The tract from the subcutaneous sphincter complex through the superficial and deep sphincters and through the levators is highly variable and often non-linear. The implementation of real-time MRI allows the advancement of a needle through the center of the muscles and a confident inclusion of the levators (Video 45.1).

PREOPERATIVE PREPARATION

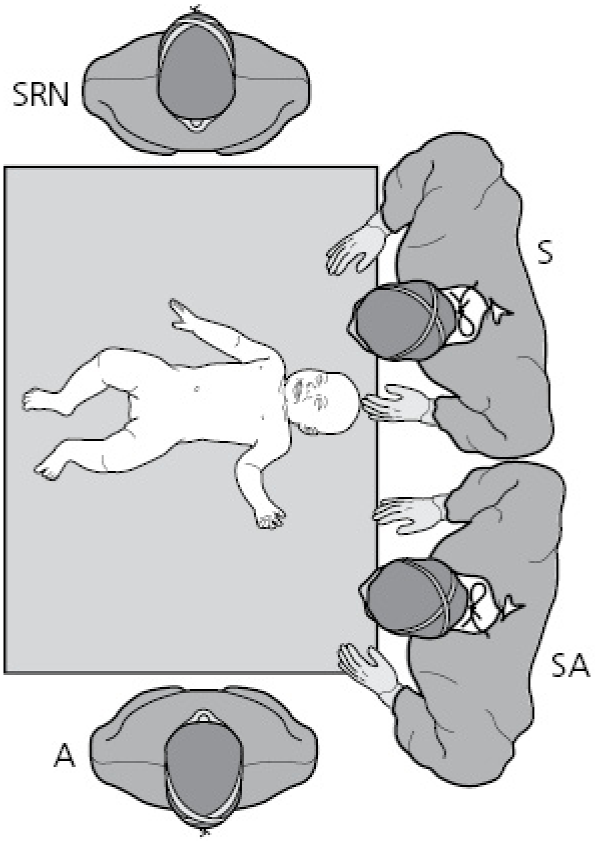
Preoperative preparation is similar to that of patients being prepared for PSARP. This is detailed in Chapter 43.

OPERATIONS

The laparoscopic repair of high ARMs can be completed in one, two, or three stages. The one-stage procedure, or primary pull-through, is performing the definitive repair without preoperative diverting or a protective ostomy. A two-stage consists of a primary pull-through with a protective ostomy and subsequent stoma takedown. In the authors' experience, the three-stage procedure is most common. This is because LAARPs are typically performed for ARM without an anal opening and a colostomy is created in the newborn period either within our institution or by referring surgeons. This stoma is left in place during the repair with subsequent takedown.

One-stage procedure: Primary pull-through

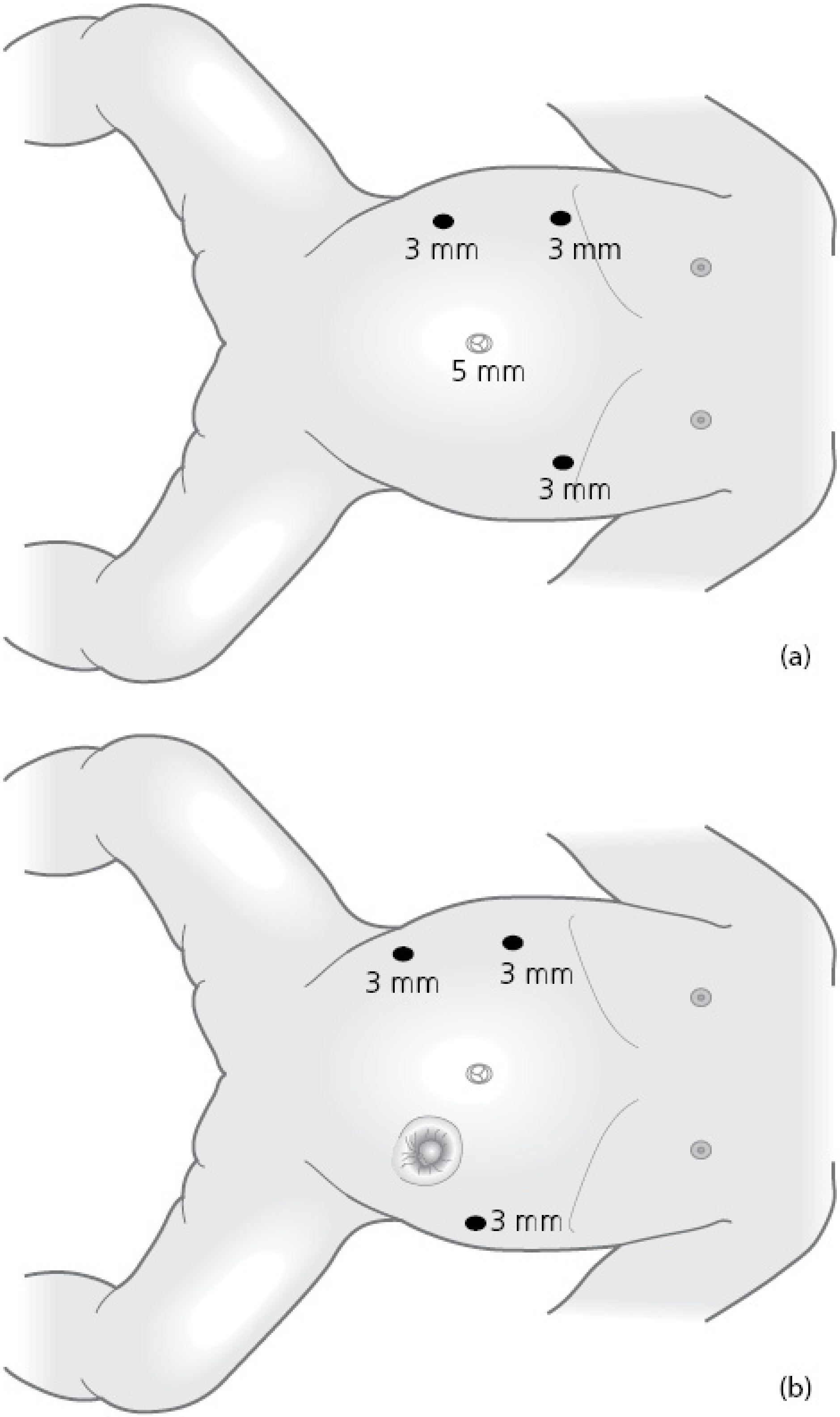
Following the institution of general anesthesia and administration of preoperative antibiotics (usually cefoxitin), cystoscopy can be performed to identify the location of the fistula and aid in the appropriate placement of a urinary catheter. The patient is positioned transversely at the foot of the operating table (**Figure 45.1**), and the child's entire torso, groin, perineum, and lower extremities are prepped. The authors use a either a split drape or extremity drape in a manner to allow the patient's mid and lower thorax, abdomen, and lower extremities into the operative field. This allows easy access to the perineum and abdomen simultaneously.



45.1

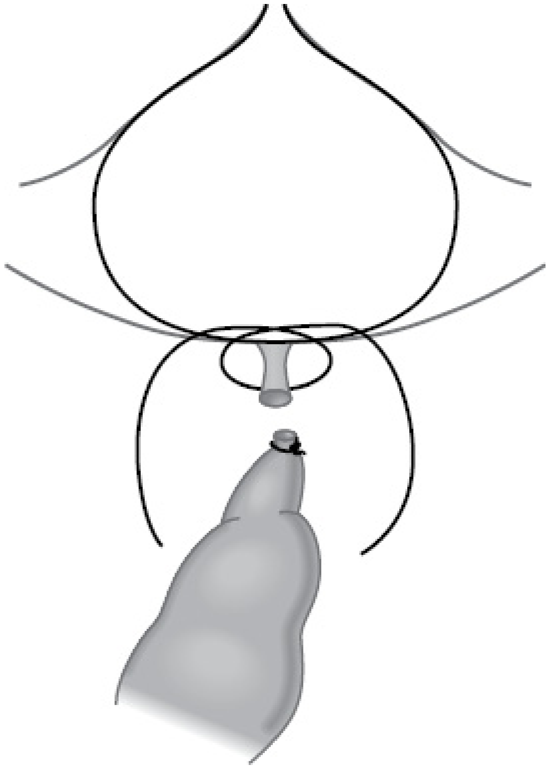
To achieve pneumoperitoneum, the authors prefer the step trocar system, but this can be left to surgeon preference. The abdomen is insufflated to a pressure of 8 cmH_2O. The pressure can then be raised to 12–15 cmH_2O to facilitate exposure.

Additional ports (3 mm or 5 mm) are placed in the left upper (or lower) quadrant, right flank, and right upper quadrant along the anterior axillary lines (**Figure 45.2a**). Care is taken that the right upper quadrant port is below the liver edge and the left sided port is optimized around the colostomy should one exist. An alternative approach is to pass the 3 mm instruments directly through the abdominal wall without trocars (**Figure 45.2b**). The table is moved to the Trendelenburg position so that the small bowel is distracted cephalad exposing the rectum and pelvis.


3 mm
3 mm
5 mm
3 mm
(a)
3 mm
3 mm
3 mm
(b)

45.2a,b

Once position, access, and exposure have been achieved, rectal dissection follows, incising the peritoneum at the peritoneal reflection with hook cautery (**Figure 45.3**). Electrocautery is then used circumferentially to dissect the mesorectum off the rectum and the dissection is carried distally, making sure to stay right on the rectal wall. If the bladder obscures the surgeon's field of vision, a stitch can be passed through the abdominal wall, through the posterior wall of the bladder and back out of the abdominal wall to temporarily suspend the bladder anteriorly. It is important to identify the ureters, vas deferens, and seminal vesicles bilaterally to avoid injury to these structures during the rectal dissection. As the rectum tapers into the fistula, it is controlled and transected leaving a short portion of the fistula tract in place. The fistula to the genitourinary tract is then closed with a 2/0 PDS□ endoscopic loop, or sutured with an absorbable laparoscopic suture. The rectogenitourinary fistula is divided at the most distal point without injuring or encroaching upon the genitourinary tract. A cystoscope can be used to guide ligation of the fistula to avoid narrowing the urethra or leaving a remnant of the original fistula. Alternatively, opening the rectal fistula to identify the position of the urethra can sometimes be helpful.

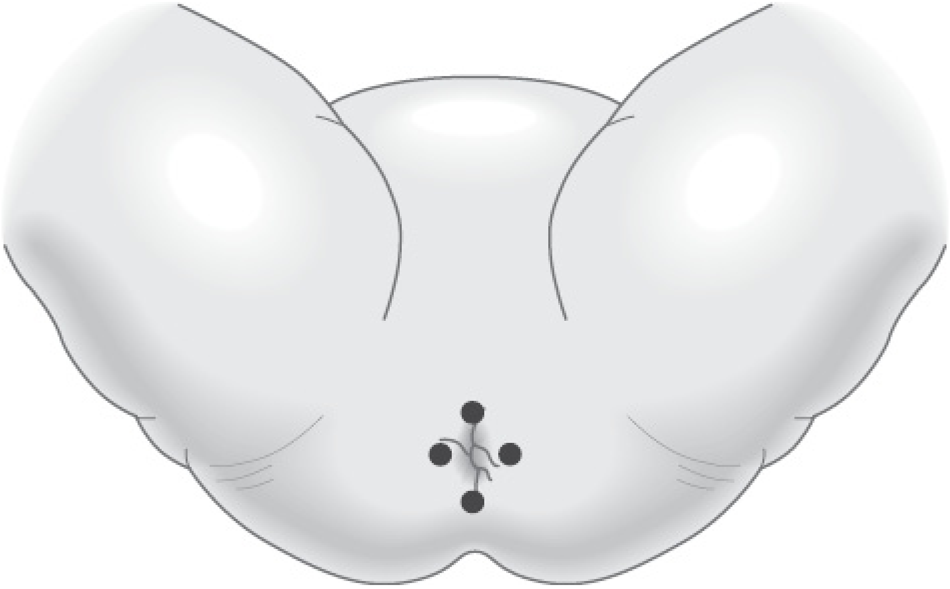


45.3

The rectum is then reflected cephalad and the pelvic floor is examined. If image-guided surgery technique is used, the previously placed MRI guided needle is identified at this point. The perineal portion is performed. In short, an incision in the marked and confirmed center of the subcutaneous sphincter complex is made through the dermis. The anal canal is created with Seldinger technique with a radially dilating trocar. The previously transected fistula is brought down and the neoanus is sewn into place (Video 45.2).

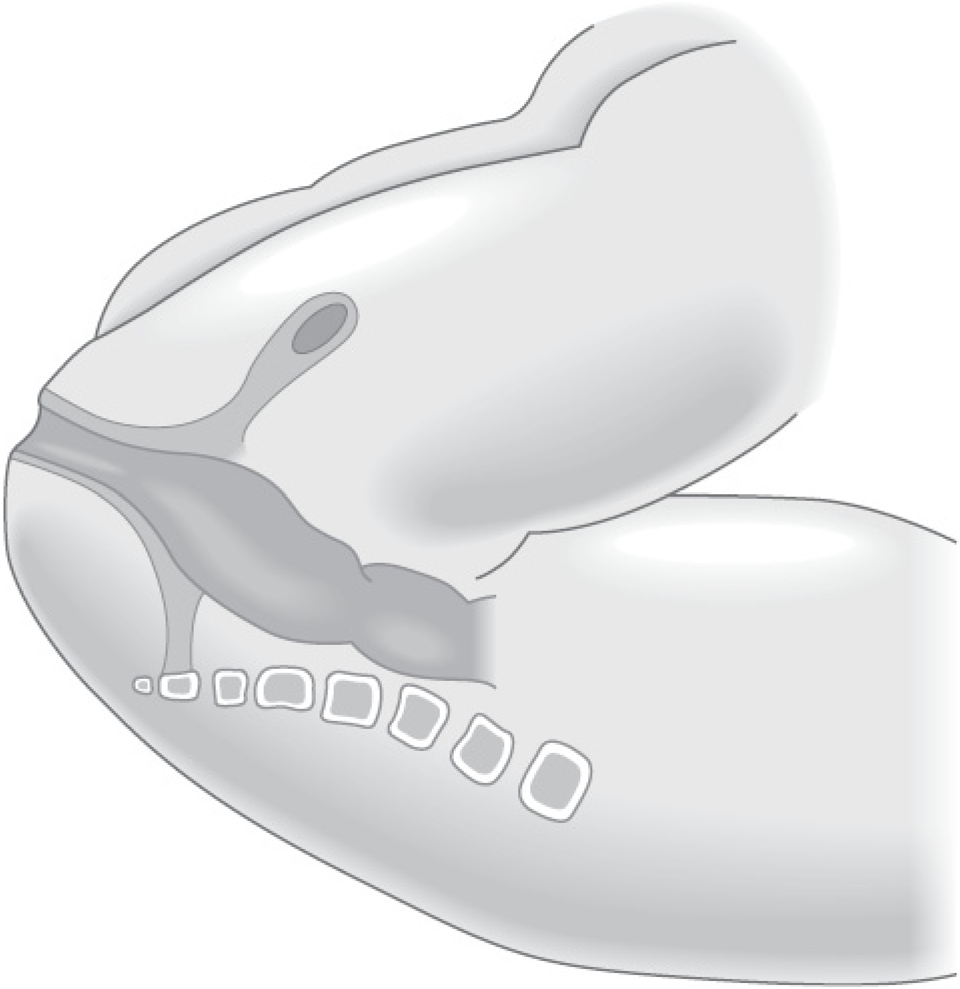
If no prior needle is in place, the space from the apex of the pubococcygeus muscle extending posteriorly is identified and developed. This will be the space through which the rectum will be passed. The vas deferens medially points to the prostate, which aids the surgeon in locating the urethra so as to avoid inadvertently injuring it. The lateral attachments of the colon may need to be mobilized in order to allow the rectum to reach the perineum.

Attention is then turned to the perineum, where the center of the superficial anal sphincter is mapped with a muscle stimulator. The area of maximal contraction is marked at its anterior and posterior limits (**Figure 45.4**).



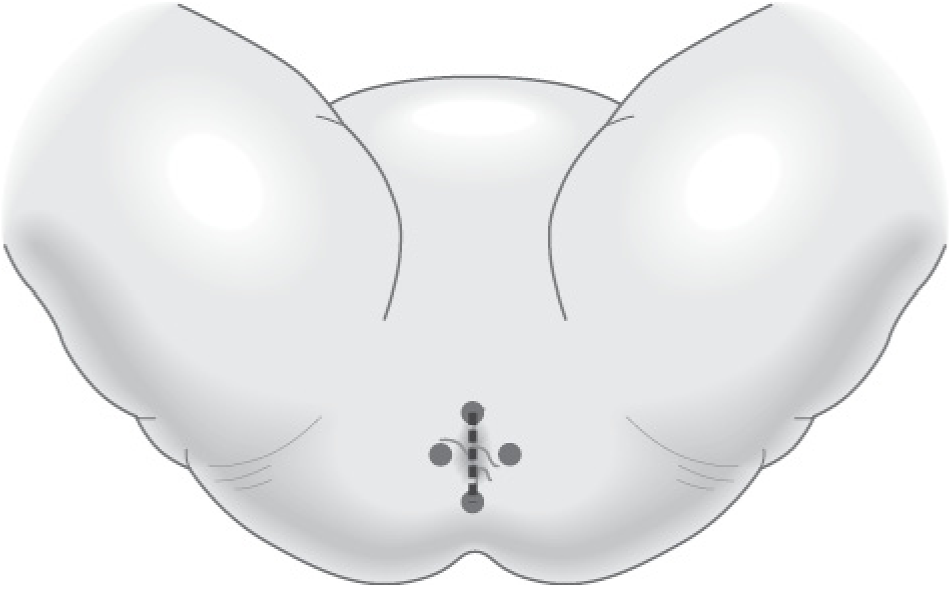
45.4

The patient's hips are flexed in such a way that the knees are directed up to the patient's shoulders (**Figure 45.5**). This position is thought to straighten the path for the pull-through and neoanus. Care must be taken as pelvic musculature can be quite variable and non-linear, and the track is best defined with the previously mentioned MR techniques.



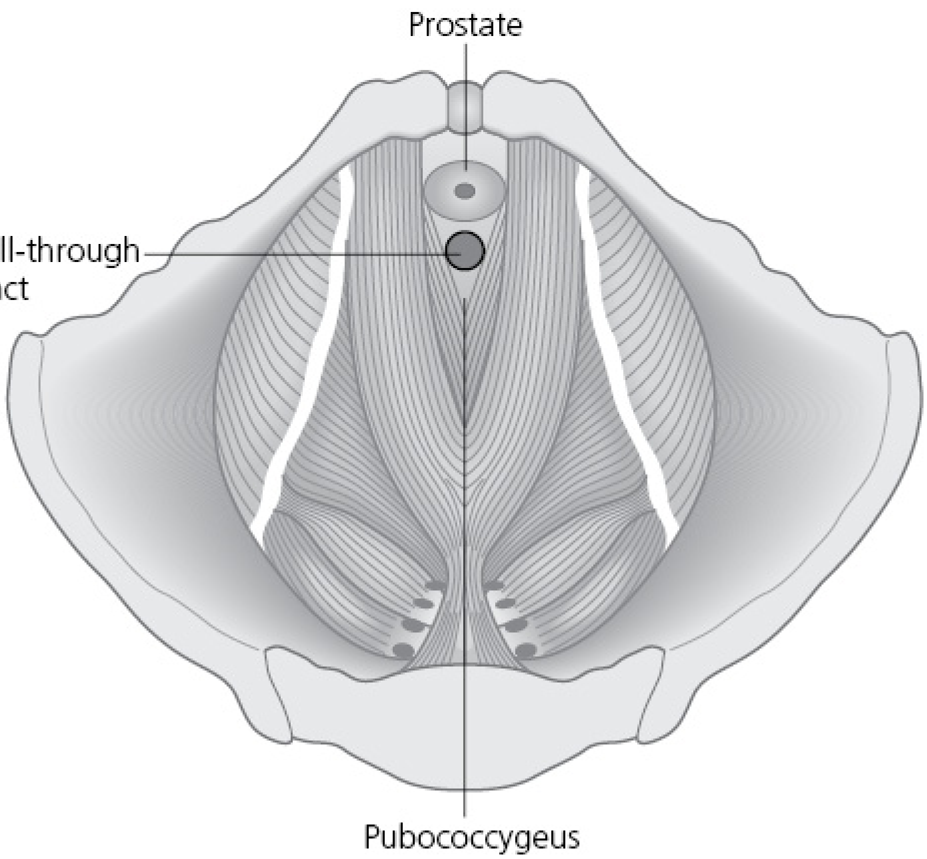
45.5

An 8 mm sagittal incision is made sharply in the center of the sphincter and the subcutaneous tissues are dissected bluntly, making every effort to stay in the midline (**Figure 45.6**). The laparoscopic transillumination is then seen at the site of perineal dissection and a 25-gauge long needle is used to identify the correct tract for the pull-through, visualizing the needle laparoscopically from above.



45.6

Next, a radially dilating trocar is employed to create the passage for the pull-through. The Veress needle and sheath are inserted under laparoscopic visualization, in exactly the same axis that the finder needle had taken, pointing toward the apex of the “V” of the pubococcygeus muscle, staying posterior to the urethra (**Figure 45.7**). The sheath is then dilated to a 12 mm port using the blunt trocars. A laparoscopic Babcock instrument is inserted through the perineal 12 mm port and the distal end of the rectum is grasped. The 12 mm perineal port, grasper, and rectum are gently pulled caudally, delivering the rectum through the sphincter complex to the perineal skin. The anastomosis is then fashioned with 4/0 Vicryl interrupted sutures. The neoanus is calibrated with Hegar dilators and the size is recorded.



45.7

The abdominal cavity and pelvis are inspected laparoscopically for hemostasis, the pneumoperitoneum is released, and all the ports are removed. The linea alba at the level of the umbilical port is reapproximated with 2/0 Vicryl® figure-of-eight suture and all the abdominal skin incisions are closed with a single Monocryl® dermal suture or skin glue. Steri-strips are applied as a dressing to the abdominal incisions.

Two-stage procedure

The two-stage procedure involves neonatal repair and creation of a neoanus as described in the section "One-stage procedure," followed by the creation of a protecting descending colostomy. The second stage in this instance is closure of the colostomy.

Three-stage procedure

A three-stage repair involves a preliminary colostomy performed in the neonatal period, followed by a pull-through several months later, and then closure of the colostomy as a third stage. There are several reasons for advocating a preliminary colostomy:

- The colon is often very dilated and full of meconium, which makes it difficult to dissect.
- The creation of a stoma permits a contrast study to be done which can identify the location of the fistula to the urinary tract.
- Some surgeons prefer to do the pull-through when the child is larger and the anatomy may be clearer and the dissection easier.

The colostomy is fashioned once the diagnosis of a high ARM is confirmed, usually approximately 24 hours after birth. The colostomy may be done in the sigmoid, descending, or transverse colon, and can be a loop or a divided stoma. We prefer to do a divided stoma in the proximal sigmoid colon. Division of the stoma prevents spillover of stool with resultant dilatation of the distal rectum. The stoma must be made proximal enough that there will be adequate length to do the pull-through in the case of a proximal fistula to the urinary tract. The colostomy can be done in the left lower quadrant, in the left upper quadrant, or through the umbilicus, and using a small incision or with a laparoscopic technique.

A thorough treatise on colostomy creation is available as a separate chapter in this volume (Chapter 51).

The second stage consists of the LAARP as described previously, without closure of the colostomy which is performed 4–6 weeks following the anoplasty. A distal contrast study through the colostomy or mucous fistula, and a simultaneous uretherogram, should be performed preoperatively to define the anatomy of the fistula.

The third stage consists of closure of the colostomy, also detailed in Chapter 51. The colostomy closure is usually performed once the anus has been calibrated to the appropriate size, which usually requires an additional 4–6 weeks following the pull-through. A distal contrast study can be done prior to colostomy closure to ensure that there is no leak or recurrent fistula.

POSTOPERATIVE CARE

Following definitive repair and creation of the neoanus, the urinary catheter is kept *in situ* for up to 5 days, depending on surgeon preference. The authors do not routinely obtain a vesicourethrogram before removing the catheter. If the catheter is inadvertently removed early, the patient is monitored for spontaneous passage of urine. The catheter should not be replaced blindly. If it becomes necessary to replace the catheter, this should be done under radiological guidance or with the aid of a pediatric urologist. The patient is maintained with IV fluids and bowel rest and is initiated on an oral diet once bowel function resumes. Postoperative antibiotics are not routinely administered after 24 hours. The patient's perineum is monitored closely for perineal sepsis and the patient remains strictly nil per rectum for approximately 2 weeks. Furthermore, when the patient's perineum is exposed to feces for the first time, it is at high risk for contact dermatitis, which can be prevented with routine liberal application of a zinc-based protective barrier cream to the neoanus and buttocks. Once the patient has resumed a full oral diet and is having bowel movements, or ostomy output, without difficulty, the patient is discharged and is seen again in 2 weeks where the neoanus is calibrated. Dilations are left to the preference of the surgeon and can be performed in the surgeon's office or by the parents at home. Many surgeons instruct the parents to dilate the neoanus on a daily basis, and others do not dilate at all until the child proves to have a stricture. Alternatively, the strictured neoanus can be repaired with a stricturoplasty at the time of colostomy takedown. Neoanus size is directly dependent on age (**Table 45.1**). Once the desired neoanus size and compliance is achieved, the dilatations are weaned.

Table 45.1 Age and Hegar size of neoanus

Age	Hegar size (mm diameter)
0–4 months	12
5–8 months	13
9–12 months	14
13–36 months	15
3–12 years	16
>12 years	17

FURTHER READING

Georgeson KE, Inge TH, Albanese CT. Laparoscopically assisted anorectal pull-through for high imperforate anus: A new technique. Presented at the 1999 Annual Meeting of the Section on Surgery of the American Academy of Pediatrics, Washington, DC, October 8–10, 1999. https://doi.org/10.1053/jpsu.2000.6925

Hsu Y, Fenner DE, Weadock WJ, DeLancey JO. Magnetic resonance imaging and 3-dimensional analysis of external anal sphincter anatomy. *Obstet Gynecol.* 2005 Dec; 106(6): 1259–65. doi: 10.1097/01.AOG.0000189084.82449.fc.

Jarboe MD, Ladino-Torres M, Wild L, et al. Imaged-guided and muscle sparing laparoscopic anorectoplasty using real-time magnetic resonance imaging. PMID: 32638078 DOI: 10.1007/s00383-020-04700-5.

46

The Malone antegrade continence enema

HENRIK STEINBRECHER

HISTORY AND BACKGROUND

The technique of the antegrade continence enema (ACE) procedure was conceived by Padraig Malone et al. working at Great Ormond Street Hospital in London in the 1980s as a treatment for intractable constipation unresponsive to any forms of non-surgical methods. It has been successfully used in thousands of patients all over the world and there are now extensive follow-up data over many years' duration.

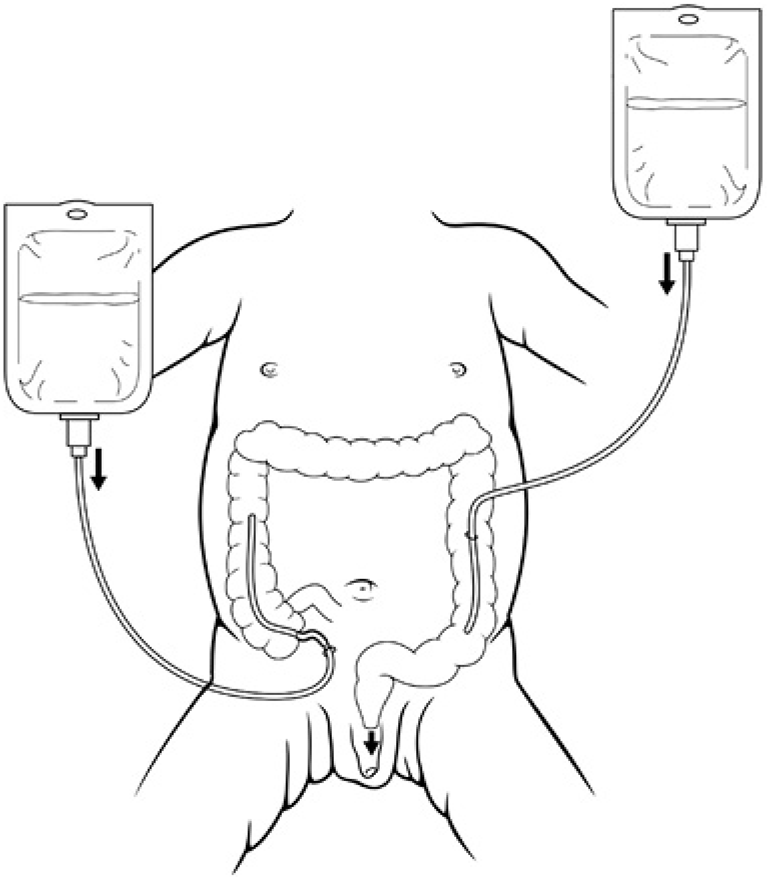
The operative details and modifications of the procedure have evolved over the years from the original disconnected reversed appendix to disconnected unreversed, undisconnected imbricated, laparoscopic antireflux/base-plicated, laparoscopic unplicated, split appendix (in conjunction with a Mirofanoff conduit), laparoscopic-assisted, or left-sided ACE. Occasionally, an endoscopic (laparoscopic-assisted) tube placement is carried out prior to formal ACE formation to assess the potential success of the antegrade washout mechanism.

Where the appendix cannot be used or is absent, conduit options include a Yang–Monti tube or a cecal tube using similar principles.

PRINCIPLES AND JUSTIFICATION

The social and psychological implications of a child with unmanageable constipation and soiling are considerable. Once all medical therapy (in the forms of drugs or retrograde washouts) has been exhausted, the last resort has traditionally been formation of a colostomy and diversion.

The ACE combines the principles of the Mitrofanoff continent catheterizable channel (devised by Paul Mitrofanoff, a French urologist working in Rouen) with that of integrated washouts or colonic lavage to empty it and prevent soiling. Depending on the aetiology of the soiling, and bowel transit times and variations, the position in the colon of the ACE may be modified (**Figure 46.1**).



46.1

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

Preoperative counselling, and the motivation of the patient and parents (or carers) are the keys to success. A successful ACE washout can take up to 45–60 minutes to complete on a daily basis and appropriate toilet facilities are mandatory for this. It is helpful at times for patients to meet someone with an ACE preoperatively. A specialist nurse can facilitate this and is an essential part of the team.

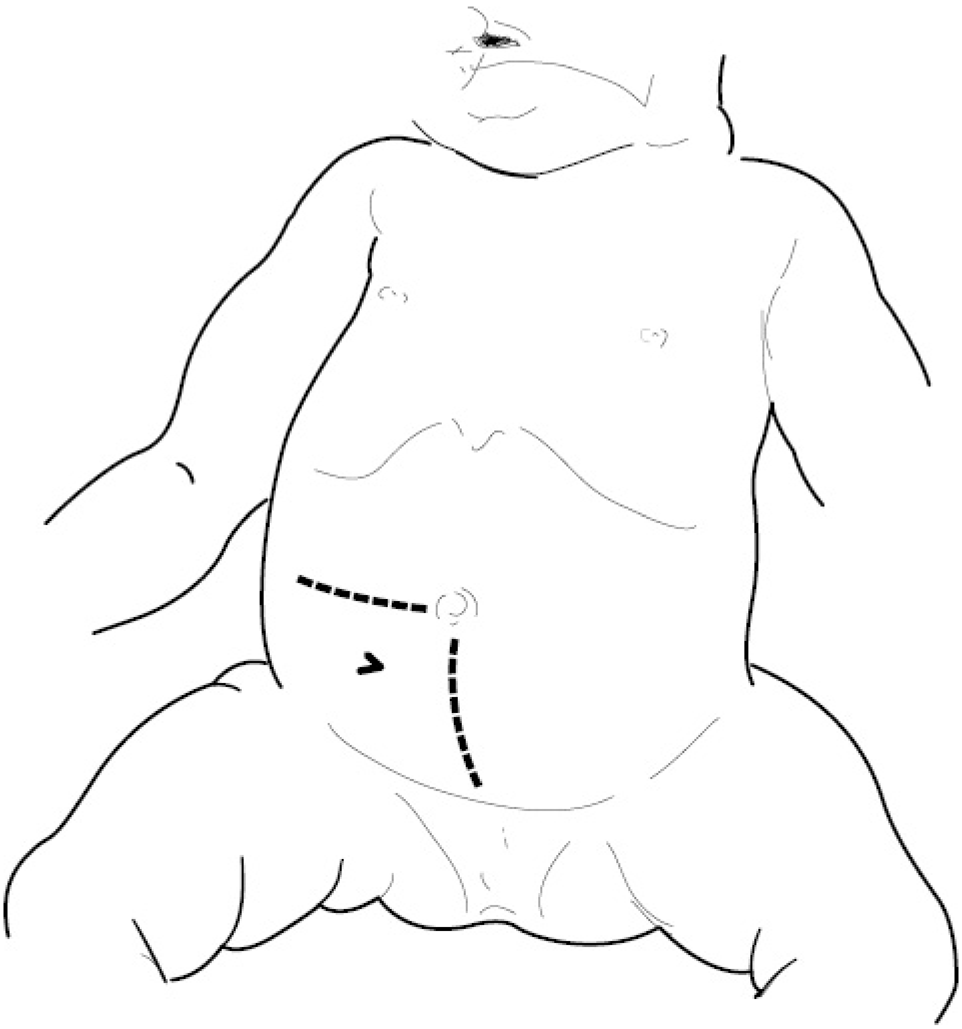
Any associated neuropathic bladder and its management must also be taken into consideration with the assistance of a paediatric urologist. This may require preoperative renal tract imaging with investigations such as a renal tract ultrasound (US) scan, dimercaptosuccinic acid (DMSA) scan, urodynamic studies, preoperative serum electrolytes and blood count, and blood cross match if major bladder reconstructive procedure is planned at the same time.

Preoperative bowel preparation is highly recommended, even in a simple isolated laparoscopic ACE as one is never sure whether other surgical maneuvers may be necessary to facilitate the ACE formation. In addition, a clear colon facilitates laparoscopy or endoscopy. Clear fluids for 24 hours prior to surgery and sodium picosulphate (e.g. Picolax™) are used in our center with a rectal washout the evening before or on the morning of surgery. Preoperative IV fluids may need to be given if the bowel preparation causes a lot of fluid loss.

Deciding on the position of the placement of the ACE conduit and stoma is influenced by the following factors: Body habitus, bowel transit time, previous surgery and scars, right- or left-handedness of the patient for catheterization, and the coexistent need for formation of a Mitrofanoff conduit for the bladder.

OPEN OPERATION

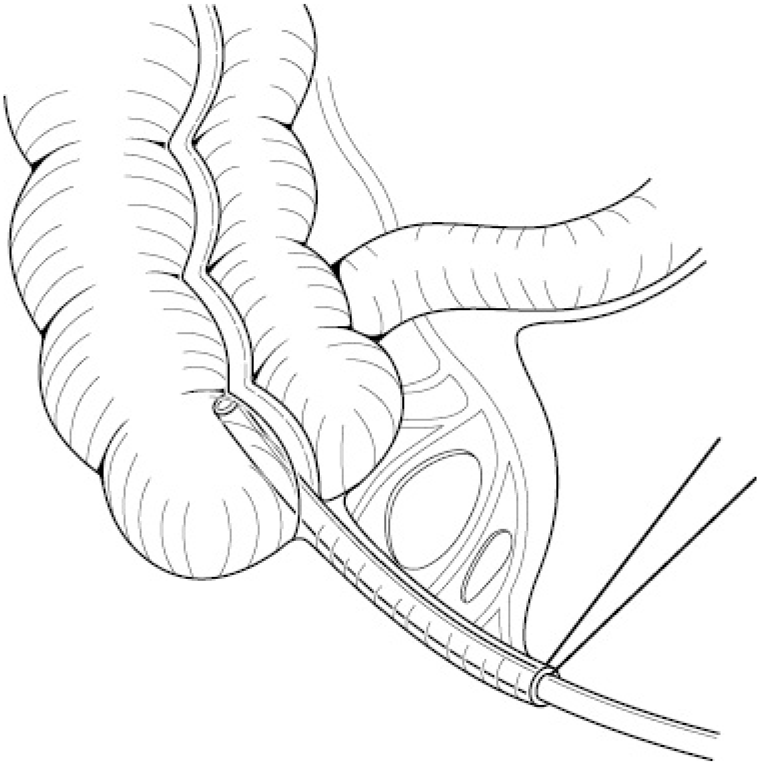
There are a number of possible incisions for this. An ACE procedure requires adequate mobilization of the cecum and ileocecal valve to allow the appendix to be moved to the location of choice for the stoma. This may require a lower midline, lower transverse, or right lower quadrant incision (**Figure 46.2**). The author favors the umbilical V stoma for an isolated right-sided ACE and uses a laparoscopic approach or lower midline incision to achieve this. In general, a laparoscopic approach is now recommended even if an antireflux maneuver (e.g. base of appendix fundoplication) is preferred.



46.2

Mobilization of cecum

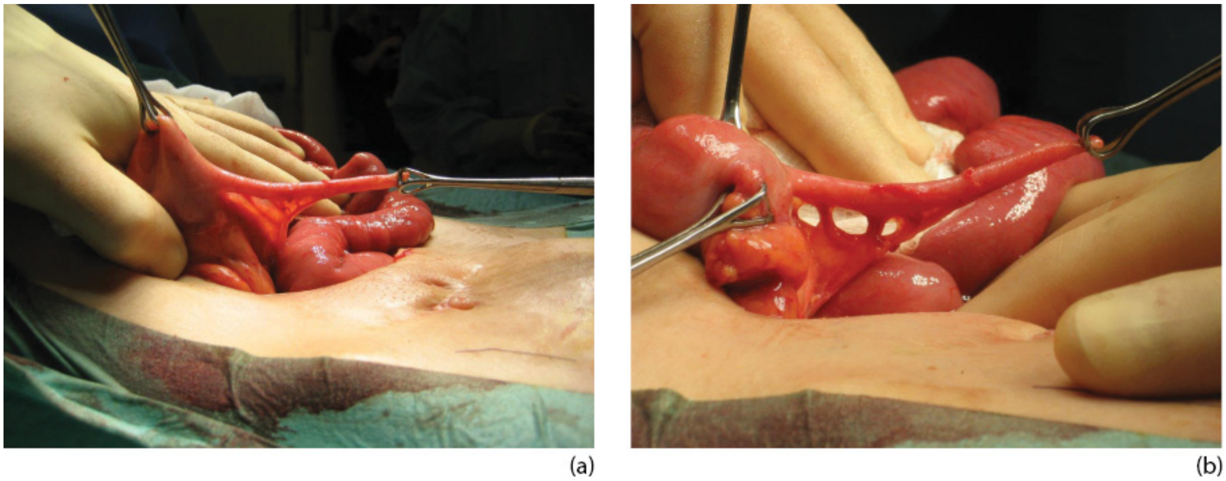
The cecum needs to be mobilized enough to be able to get the appendix base to the site of the proposed stoma and to allow adequate freeing of adhesions that may be a cause of kinking (**Figure 46.3**).



46.3

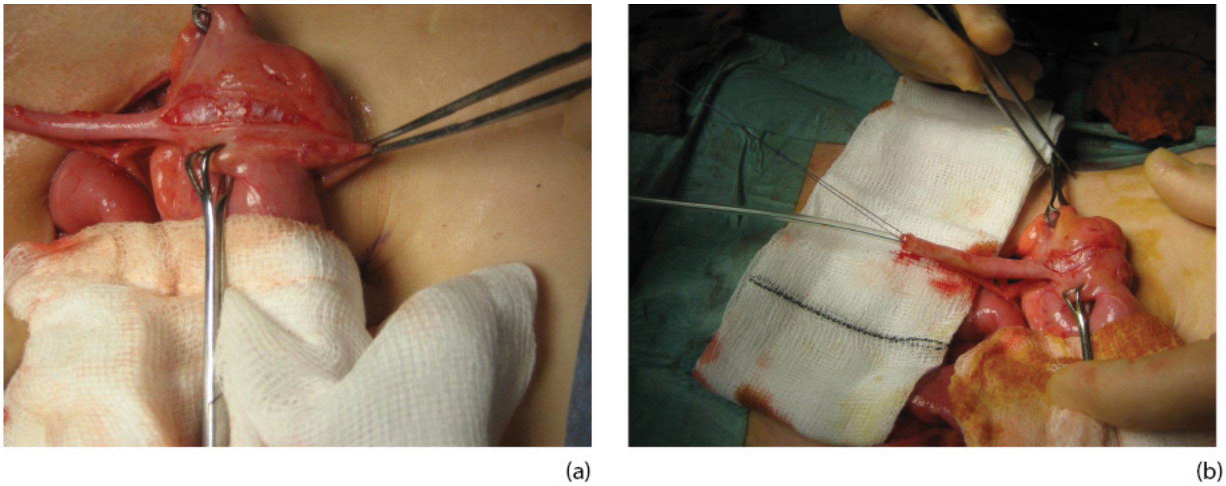
Mobilization of appendix

The appendix is mobilized on the appendicular artery ensuring good preservation of the vascular arcade (**Figure 46.4a and b**). The stump is excised after placement of a stay suture (4/0 Vicryl□) to allow it to be pulled through the stoma. The tip is sent off to histology in case of the rare event of there being an incidental carcinoid within. A 12 Fr feeding tube is passed through the appendix ensuring its luminal patency.



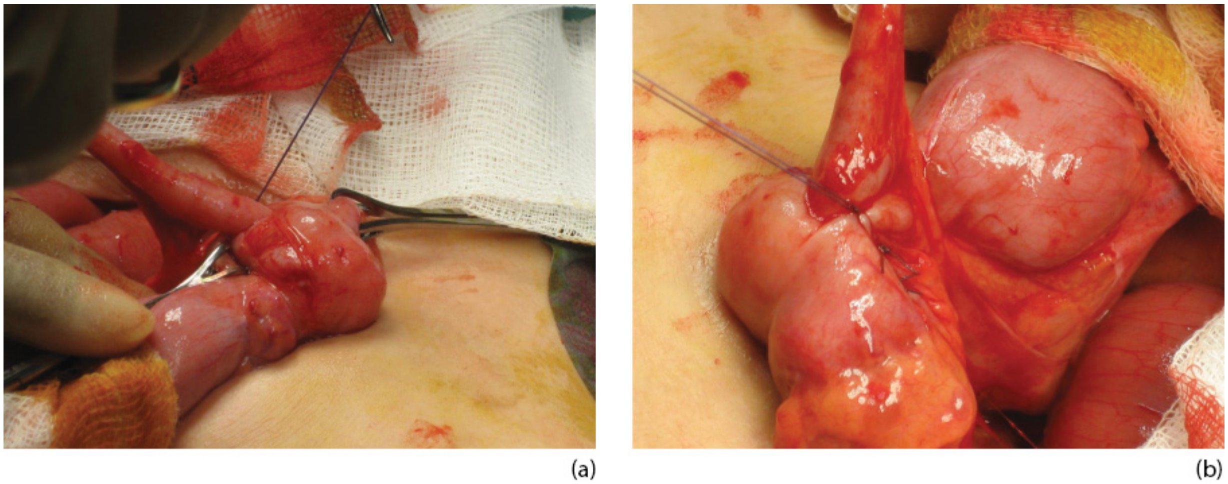
46.4a,b

For an imbricated appendix ACE, mesenteric windows need to be created carefully without injury. The teniae coli are identified and an appropriate one, usually the middle of the three, is incised carefully from the base of appendix for about 4–5 cm (**Figure 46.5a and b**). The edges of the tenia are then split — akin to splitting a pylorus — allowing the mucosa to protrude freely. A "V"-shaped split is then created in the tenia at appendix base level to allow the latter to be folded without obstruction once the appendix has been laid into the trough.

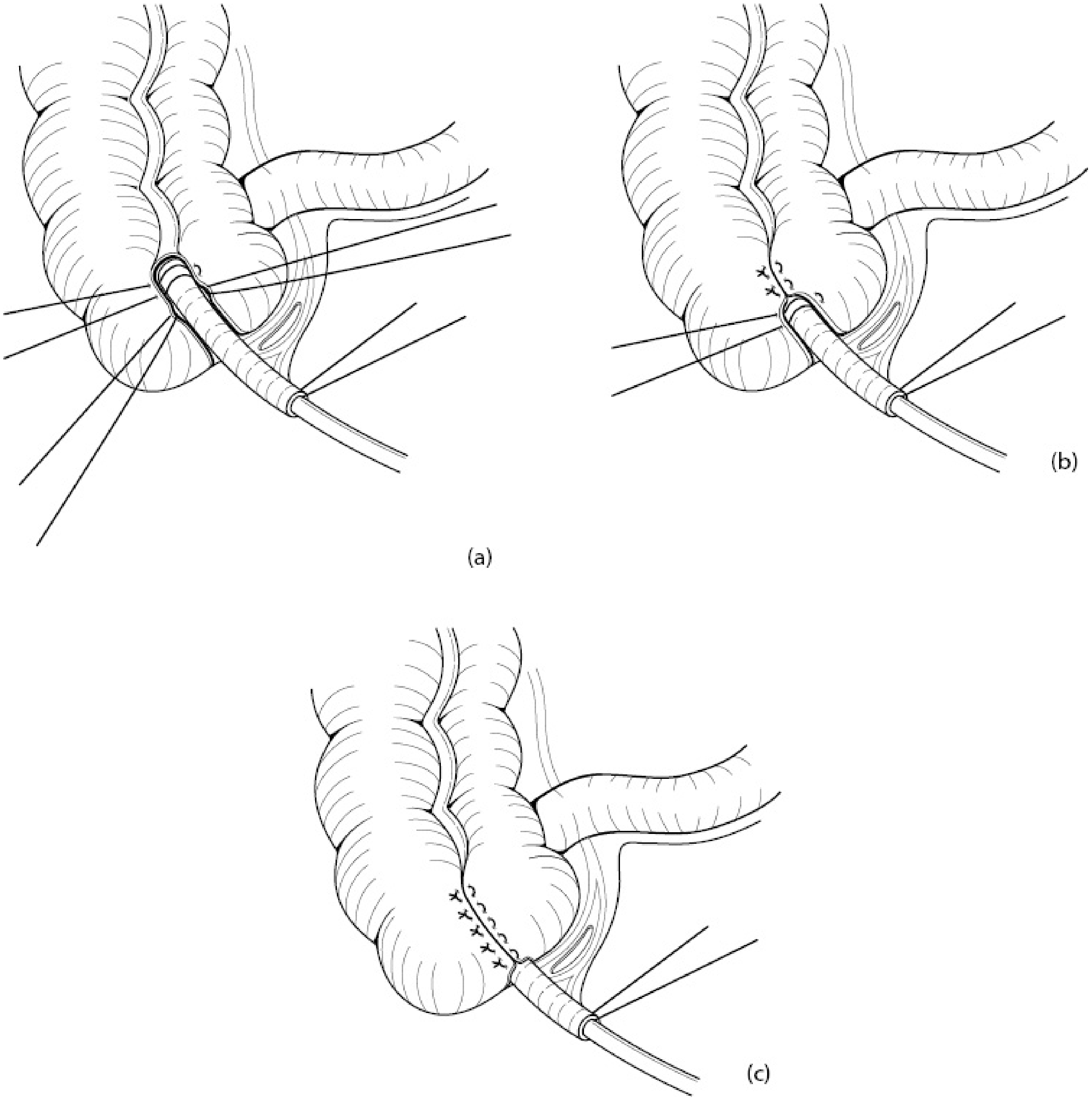


46.5a,b

4/0 Vicryl® sutures are then placed from one tenia and cecal wall to the other through the mesenteric fenestration such that the appendix is buried in the trough wrap with the mesentery uppermost and the sutures tied through the fenestration gaps (**Figure 46.6a and b**). This conserves the blood supply and buries the appendix into a submucosal tunnel (**Figure 46.7a–c**).



46.6a,b

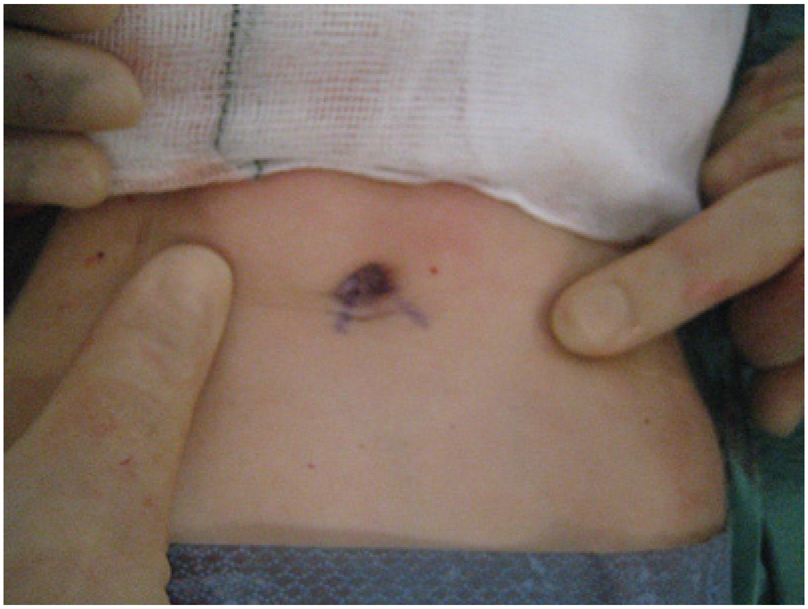


46.7a–c

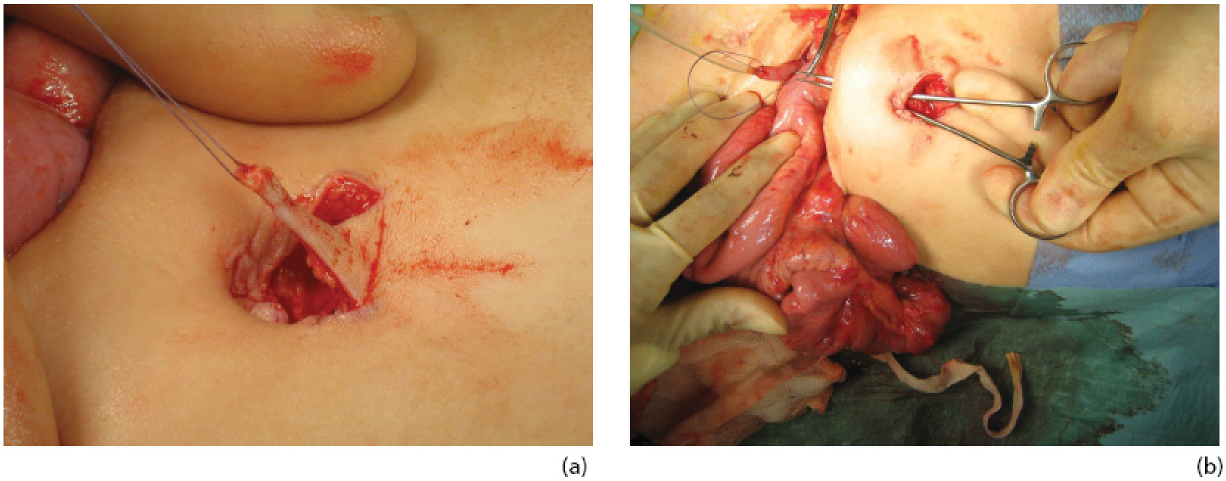
The tunnel is completed so that about 1 cm of appendix, or longer if the abdominal wall is thicker, is left unwrapped (hence the need to get the base of appendix to stoma level on mobilizing the cecum). This short free element of the appendix is then brought through the stoma site. The cecum is fixed to the back of the abdominal wall using three 4/0 Vicryl® sutures.

Stoma creation

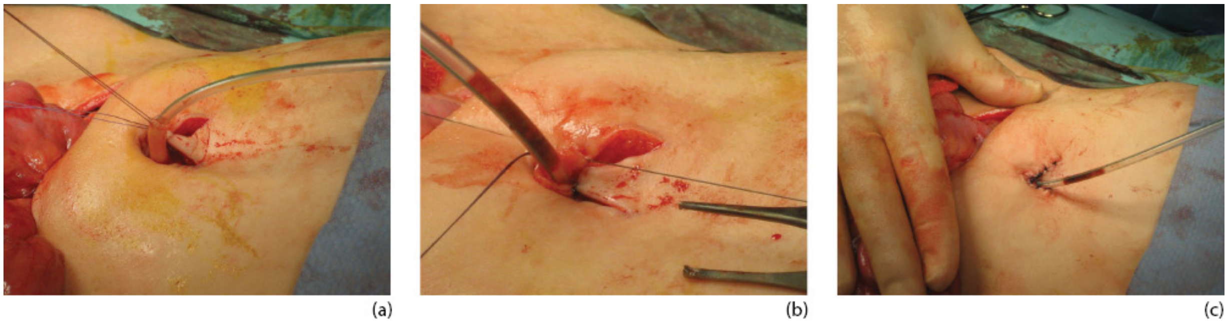
The stoma is created in a number of ways depending on its site. At the umbilicus a V shaped skin flap is created (**Figure 46.8**). The umbilicus is dissected down through the abdominal wall and a hole is created in the peritoneum that will admit the appendix tip containing the 12 Fr feeding tube (**Figure 46.9a and b**). The appendix edge is fish-mouthed and the V flap is inserted and anastomosed using interrupted 5/0 Maxon™ suture with a tapered needle (which passes through the thick umbilical skin more easily) (**Figure 46.10a–c**).



46.8

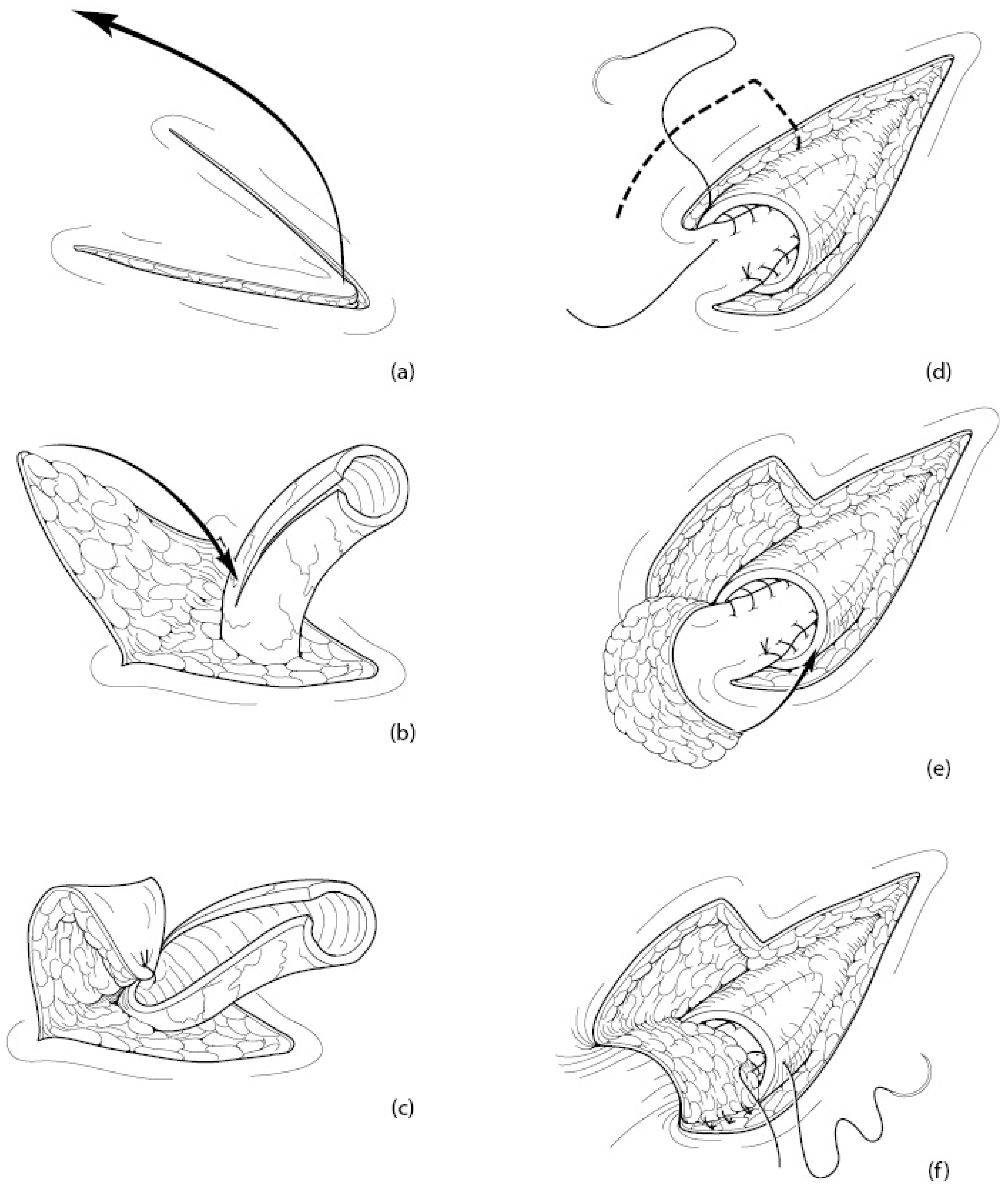


46.9a,b

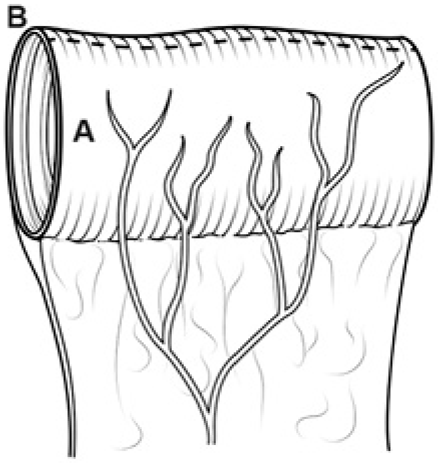


46.10a–c

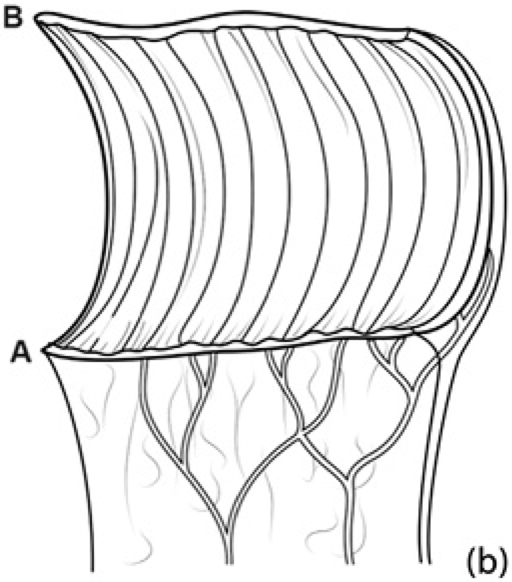
For a stoma away from the umbilicus, VQZ skin flap configuration is used with the same principle of inserting a skin flap into a fish-mouthed appendix and then bringing skin flaps over the top of the appendicular serial surface to ensure the mucosal skin interface is buried (**Figure 46.11a–f**).



46.11a–f “VQZ” flap. (a) “V” incision; (b) “Q” - small side incision (c) inset into fish-mouth of appendix; (d, e and f) finished appearance resembles a “Z”.



(a)



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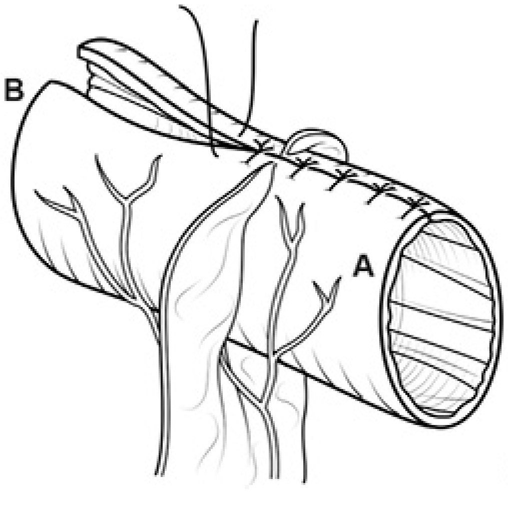


(c)

46.12a–c Yang-Monti principle. (a) antimesenteric incision; (b) display of valvulae conniventes and (c) 90 degree re-orientation and reconstruction of tube.

Absent appendix or left-sided ACE: Yang–Monti procedure

When the appendix is absent or unsuitable (too short, incomplete lumen, previously removed), or a left-sided ACE is required, the Yang–Monti method of creating a catheterizable channel can be employed.

A length measuring about 2 cm of ileum is isolated on its blood supply, divided longitudinally along its antimesenteric border, and then reconstructed at 90 degrees to its axis to align with the valvulae conniventes to create a length (2π × radius) of catheterizable channel. Interrupted 5/0 PDS® sutures are used over a 12 Fr feeding tube. The creation of a subtenial tunnel in the cecum is as described above. A small artery forceps is used to make a hole in the exposed cecal mucosa and the Monti channel is then anastomosed using interrupted 5/0 PDS® in an end-to-side manner with the feeding tube *in situ*. The other end of the Monti is brought through the stoma site. It is difficult to imbricate/plicate the Monti like the appendix due to its precarious blood supply, and the cecal tunnel is closed with interrupted 4/0 Vicryl®, incorporating the serial wall of the Monti each time to prevent the channel moving out of the tunnel.

Double Montis (two Montis anastomosed together), eccentrically incised Montis (where the split is off-center providing the blood supply source mainly toward one end of the Monti), or spiral Montis have all been used in formation of a Mitrofanoff channel and can also be used for creation of an ACE if needed. Stomal and catheterizable problems do, however, increase with each modification and the best long-term conduit is still the appendix.

Split appendix

When an ACE is required in conjunction with a Mitrofanoff, it is not unusual to be able to use the appendix for both rather than the appendix for one and a Monti for the other. Essential requirements to be able to use a split appendix are adequately long and mobile appendix, long appendicular artery and mesentery, mobile cecum, and mobile bladder augmentation to allow both conduits to reach their respective stomas with no tension on either blood supply. In this situation the author prefers to use the proximal appendix *in situ* for the ACE to the umbilicus and the distal Mitrofanoff channel to a right lower quadrant. The cecal end of the appendix can occasionally be elongated by tubularizing part of the most dependent part of the cecum, although this leaves a longer bowel suture line at that level with associated risks of leakage.

LAPAROSCOPIC OPERATION

Port placement

Laparoscopic surgery has made the creation of an ACE quicker and easier and has allowed results of non-imbricated and non-plicated ACEs to be assessed. The majority of surgeons now just “pull the appendix out of the stoma” without any other associated antireflux maneuvers.

Two or three ports are placed (5 mm or 3 mm), one in the umbilicus (for the camera) after first creating the V skin flap, and one or two others in either the right upper or right lower quadrant, and left lower quadrant. This allows good triangulation and mobilization of the cecum and appendix adequately. Occasionally, a fourth port is placed suprapubically. The appendix is mobilized fully. preserving its blood supply. and the camera is then moved to a non-umbilical port to allow the appendix to be pulled through the umbilicus. The tip is excised, a feeding tube is passed, and the stoma is sutured as before. Internal hernias and reflux have not proved to be problematic, although minor degrees of reflux do occur. The longer the appendix, the less likely is reflux.

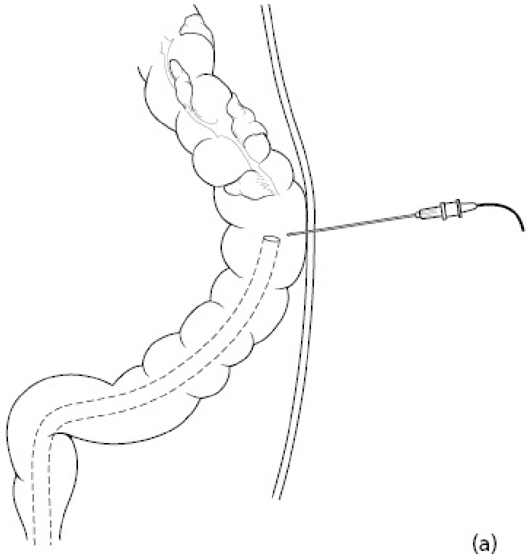
LAPAROSCOPIC-ASSISTED OPERATION

There are occasional situations when a laparoscopic-assisted ACE procedure is carried out (e.g. joint ACE–Mitrofanoff procedure, split appendix ACE–Mitrofanoff, placement of ACE stoma in non-routine position, part of laparoscopic augmentation cystoplasty).

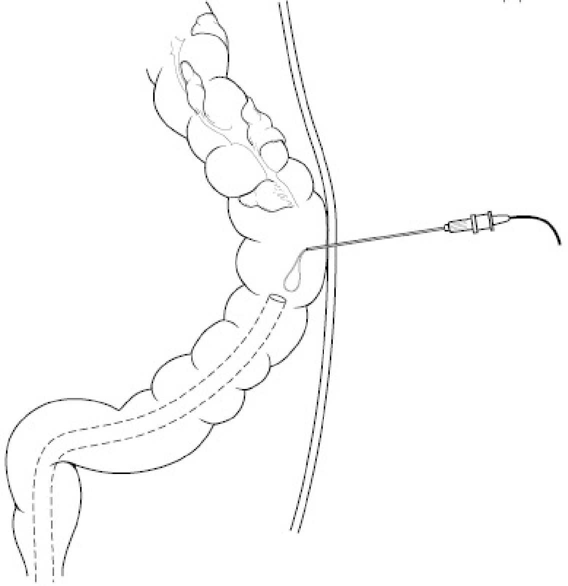
The aims in this situation are to carry out some of the mobilization and dissection laparoscopically, to place the stoma in an appropriate site on the abdominal wall with the mobilized appendix/cecum beneath, and to reduce the size of the ultimate open incision, thus improving recovery.

LAPAROSCOPIC-ASSISTED–COLONOSCOPIC PLACEMENT OF ANTEGRADE WASHOUT TUBE

In some instances it is not clear where best to site the ACE and it may be useful to place a percutaneous tube first, either in the descending or transverse colon, using the colonoscope (**Figure 46.13a–e**). A laparoscope may be used to assist more accurate placement of the tube in the appropriate part of the colon and to avoid perforation of other loops of bowel. In this combined approach, it is generally better to place the laparoscope first with one or two other instruments (often via direct port insertion into the abdomen under laparoscopic vision before the gastroenterologist places the colonoscope as inflation of the colon will make laparoscopy more difficult). With a good combined team approach, a laparoscopist may be able to guide the colon over the colonoscope without the need for much colonic insufflation. Subsequently, a Seldinger wire technique is used to place the colonoscopy washout tube.



(a)



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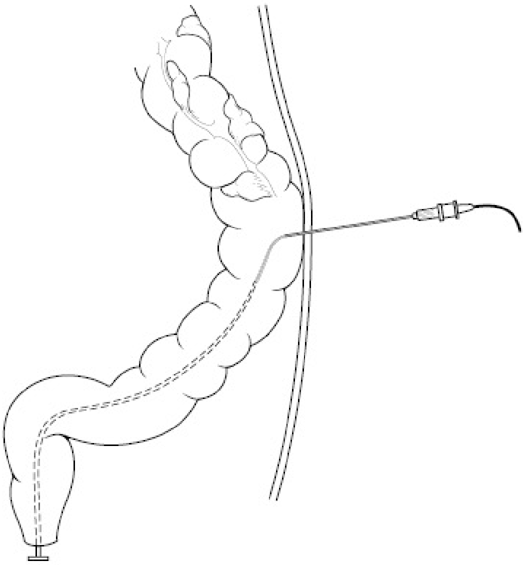

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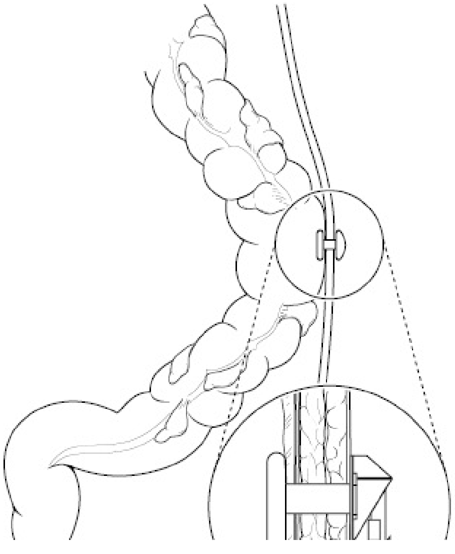


46.13a–e

POSTOPERATIVE CARE

The first use of the ACE is carried out after about 72 hours, with a tap water or saline lavage of about 5–10 mL/kg. Subsequently, one of a number of differing regimes can be used, including phosphate and water, liquorice and water, or water alone as a 20 mL/kg/washout. The washouts are carried out with an indwelling catheter in the stoma for 4 weeks, when the patient returns to learn intermittent catheterization. Once this is established, washout regimes are usually carried out every 1–2 days, depending on the soiling-free period obtained.

COMPLICATIONS

Complications may include wound infection and stomal problems (e.g. leakage, creation of a false passage with repeated catheterizations, stomal stenosis). More significant problems include conduit stenosis or ischemia (more likely if a Monti is used).

During the washout, some patients initially experience significant discomfort and pain and this is thought to be due to the cecal distension and contractile effect of the phosphate. It can be treated by reducing the enema fluid or stimulant used, but there is always a balance between giving an effective enema versus ameliorating the discomfort. This often abates with time but in some can lead to disuse or the need to resite the ACE further distally.

Stomal false passages are managed by formal placement under anesthetic of an indwelling catheter, occasionally using a Seldinger guidewire technique allowing the conduit false passage to heal.

Stomal stenosis will require dilation or refashioning, often using a modified V–Y plasty.

Ischemic stenosis will require reforming of a new conduit. This is rarely needed given the good blood supply of the appendix/cecum, but occasionally occurs in an isolated Monti ACE.

OUTCOME

Long-term data suggest that 30–50% of patients discontinue to use their ACE into adulthood as their bowel habits change, their preference for ante versus retrograde washouts changes, or as other techniques for bowel management develop.

VIDEOS

OPS_8e_video_46.1 Laparoscopic ACE
OPS_8e_video_46.2 Open ACE
OPS_8e_video_46.3 Percutaneous endoscopic ACE

FURTHER READING

Antao B, Ng J, Roberts J. Laparoscopic antegrade continence enema using a two-port technique. *J Laparoendosc Adv Surg Tech A* 2006; 16: 168–73.

Chan DSY, Delicata RJ. Systematic review: Meta-analysis of antegrade continence enema in adults with faecal incontinence and constipation. *Br J Surg* 2016; 103: 322–7.

Churchill BM, De Ugarte DA, Atkinson JB. Left-colon antegrade continence enema (LACE) procedure for fecal incontinence. *J Pediatr Surg* 2003; 38: 1778–80.

Curry JL, Osborne A, Malone PS. How to achieve a successful Malone antegrade continence enema. *J Pediatr Surg* 1998; 33: 138–41.

Gerharz EW, Vik V, Webb G, Woodhouse CRJ. The in situ appendix in the Malone antegrade continence enema procedure for faecal incontinence. *Br J Urol* 1997; 79: 985–6.

Malone PS, Ransley PG, Kiely EM. Preliminary report: The antegrade colonic enema. *Lancet* 1990; 336: 1217–18.

Monti PR, Lara RC, Dutra MA, de Carvalho, JR. New techniques for construction of efferent conduits based on the Mitrofanoff principle. *Urology* 1997; 49: 112–15.

Rawat DJ, Haddad M, Geoghegan N, et al. Percutaneous endoscopic colostomy of the left colon: A new technique for management of intractable constipation in children. *Gastrointest Endosc* 2004; 60: 39–43.

Webb HW, Barraza MA, Crump JM. Laparoscopic appendicostomy for management of fecal incontinence. *J Pediatr Surg* 1997; 32: 457–8.

47

Hirschsprung disease

MATTHEW W. RALLS AND ARNOLD G. CORAN

PRINCIPLES AND JUSTIFICATION

Although our understanding of the embryogenesis and surgical care of Hirschsprung disease have advanced considerably over the past few decades, significant complications continue to be associated with this process. One must maintain a high degree of suspicion for the disease. Suction rectal biopsy and anal manometry studies have allowed for easier and less invasive methods of establishing the diagnosis; however, obtaining details of the infant's history is the most important first step. Failure to pass meconium within the first 48 hours of life, complaints of constipation, and symptoms of enterocolitis should always be followed by a complete clinical evaluation for Hirschsprung disease.

The classic approach to the neonate diagnosed with Hirschsprung disease had been to perform a leveling colostomy and to wait until 6–12 months of age to perform the definitive pull-through. This approach has changed dramatically over the past three decades, and transition to primary pull-through is now predominant. Leveling colostomy is now reserved for patients presenting with a massively dilated colon or critically ill from complications of the disease, particularly in a child who has gone undiagnosed. Use of laparoscopy to facilitate both the diagnosis and the pull-through procedure has also become common practice.

FULL-THICKNESS RECTAL BIOPSY

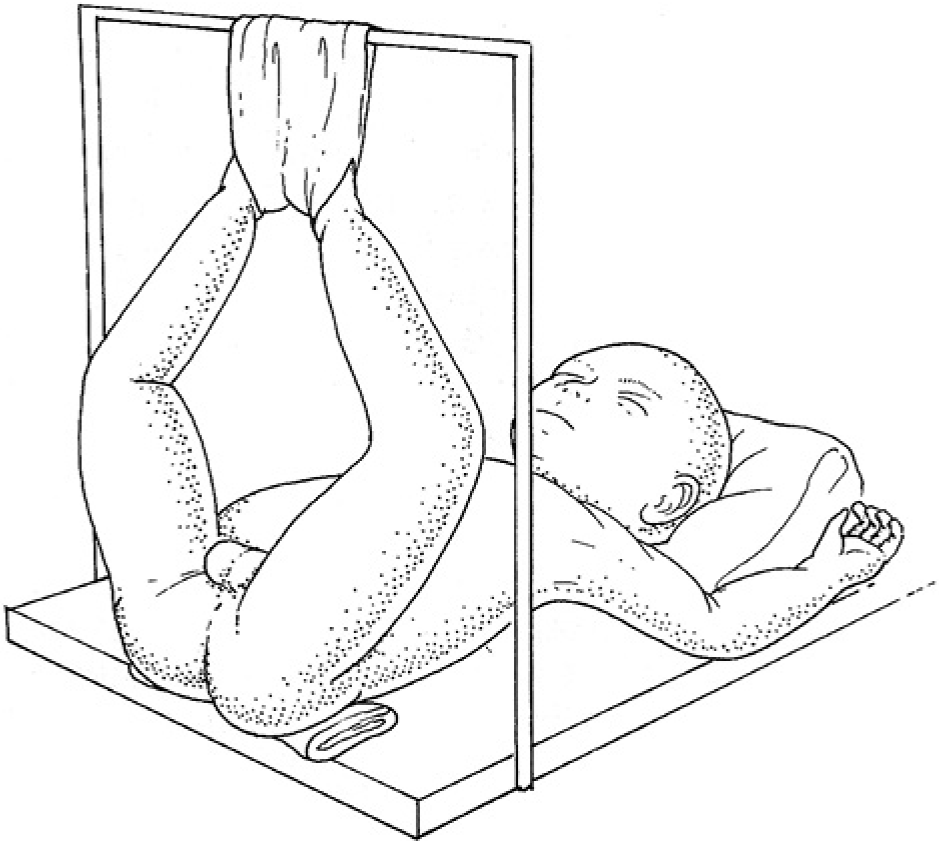
Suction rectal biopsy, because of its relative ease and low morbidity, has become the most established diagnostic tool for Hirschsprung disease. Nevertheless, full-thickness rectal biopsies are occasionally required, and the technique of full-thickness biopsy is presented here to assist surgeons who are not familiar with the procedure. Perhaps the most common indication for a full-thickness biopsy is the child who has undergone more than one indeterminate suction rectal biopsy. Another indication is the older child whose mucosa is so thick that an adequate biopsy of the submucosa cannot be obtained using a suction method.

Preoperative

No formal bowel preparation is required. The child's rectum is irrigated with saline or dilute povidone iodine solution. A sponge is placed into the proximal rectal vault and the child is placed in Trendelenburg position to prevent stool from entering the operative field. One dose of preoperative broad-spectrum antibiotics is given.

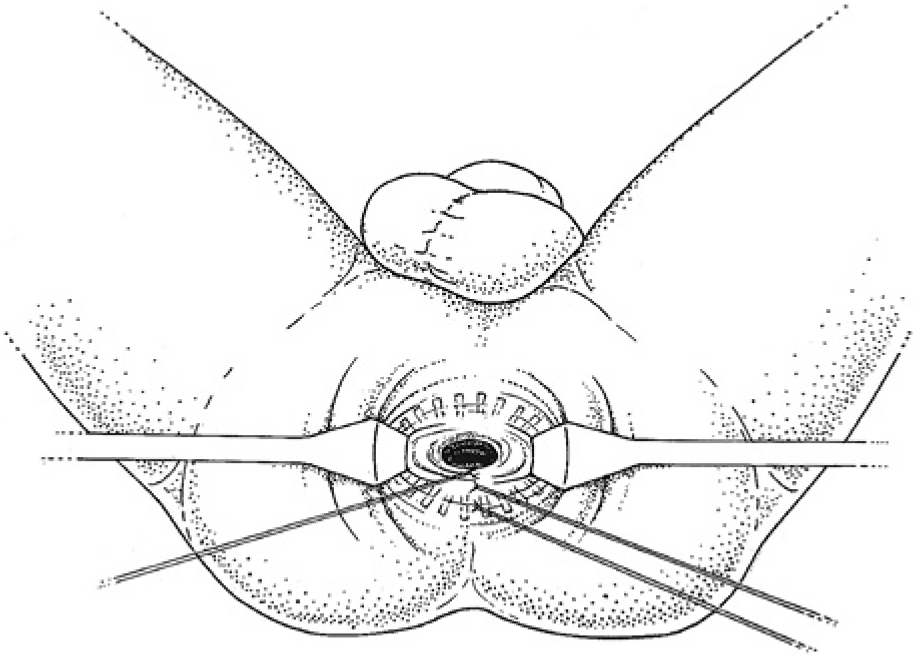
Operative technique

The patient is placed in the lithotomy position with the buttocks at the very end of the bed, supported with a folded towel. The feet are placed together (plantar surfaces adjoined) with a cotton roll, and both legs are suspended on an ether screen, or similar device (**Figure 47.1**). Flexion at the hips facilitates exposure.



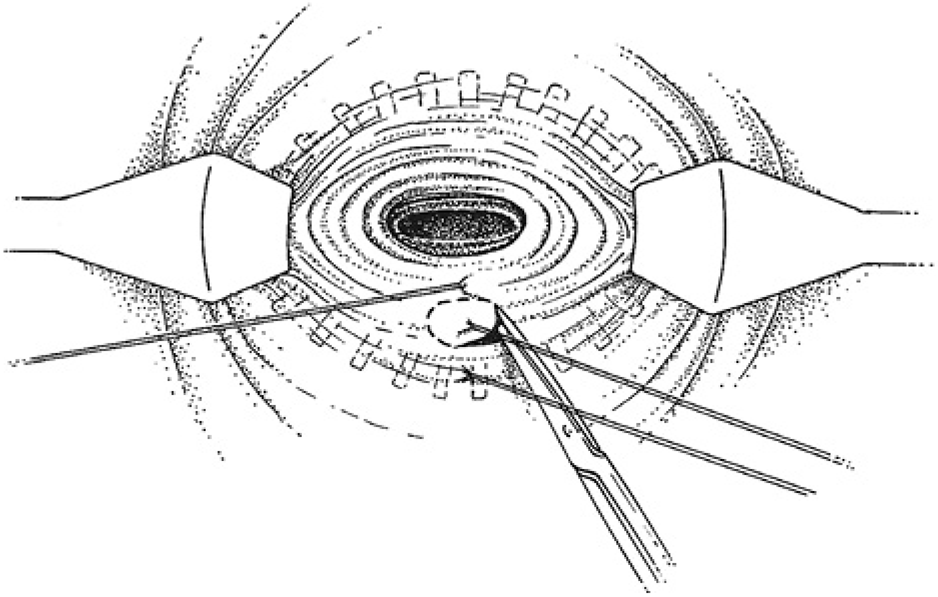
47.1

Digital dilation is followed by placement of two narrow anal retractors, a Hill-Ferguson retractor, or skin hook retractor system to enable adequate visualization (**Figure 47.2**). The superior aspect of the dentate line is identified in the posterior midline and marked with an absorbable suture (3/0), which is used for traction. Two additional polyglactin sutures are placed on the posterior wall of the rectum at 1 cm and 2 cm proximal to the dentate line. The needle is retained on the most cephalad of these sutures, as it will be used to close the defect after the biopsy is obtained.



47.2

The surgeon's non-dominant hand holds the middle suture which will retract the biopsy site. Using sharp curved scissors, a full-thickness incision is made along the lower half of the rectal wall, between the dentate line and the middle suture (**Figure 47.3**). Once this is done, the scissors can be placed in the presacral space and gently spread. Bleeding can slightly obstruct the view at this point; however, by maintaining traction on the middle suture, the upper half of the rectal biopsy is completed with two smooth cuts of the scissors, each sweeping around one-half of the tissue suspended by the middle suture. The specimen is inspected and delivered off the table. The rectal defect is closed in a single, full-thickness running or interrupted layer. This can be done with the proximal-most suture on which the needle was retained. Hemostasis is achieved fairly quickly once this suture has been placed.



47.3

LEVELING COLOSTOMY

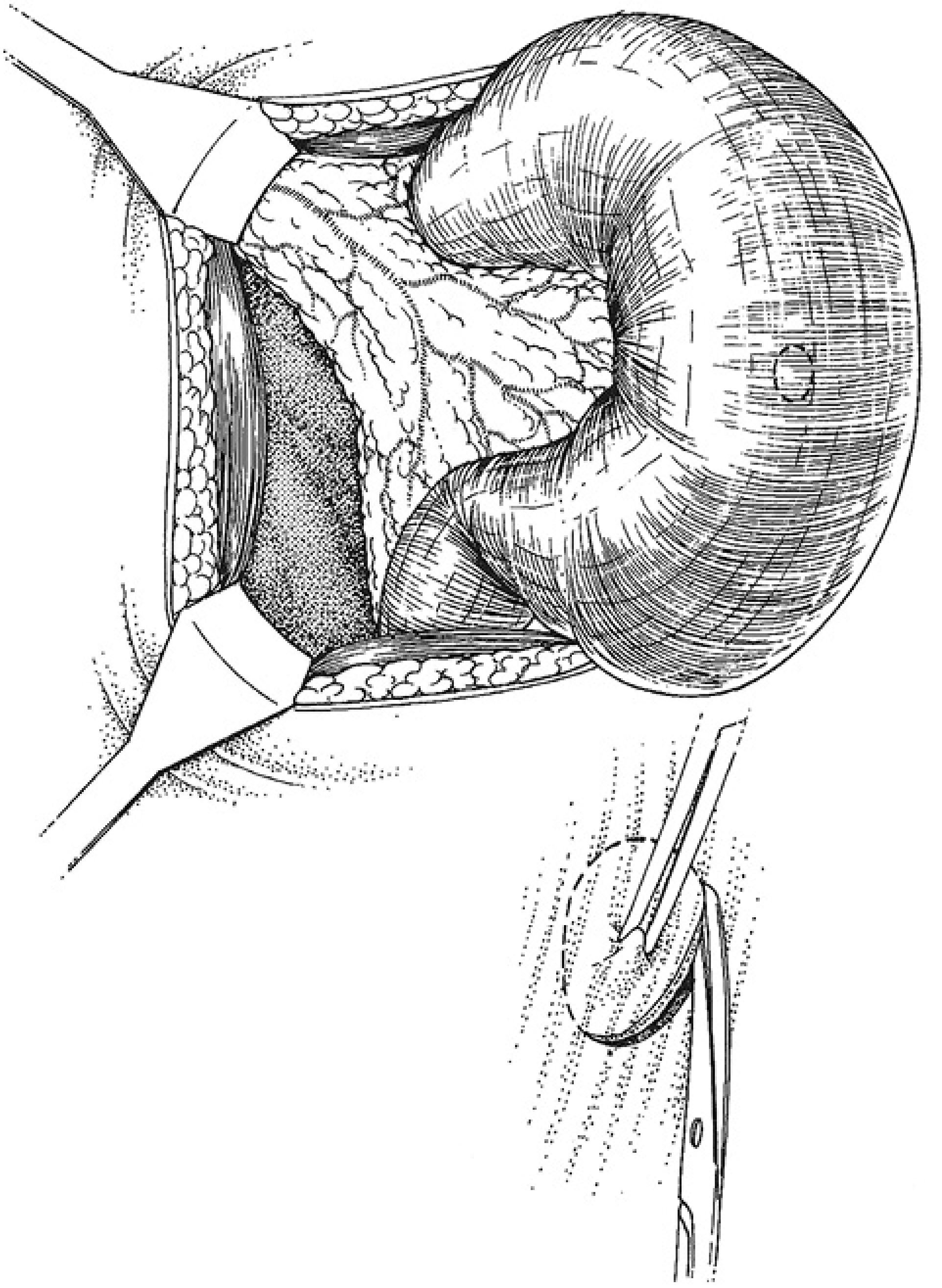
Although a primary pull-through is preferred in most infants, use of a leveling colostomy is required in infants presenting with a severe episode of enterocolitis at the time of diagnosis. A colostomy may also be required in those patients with a delayed diagnosis of Hirschsprung disease, where the colon has become overly distended and not amenable to a primary pull-through. Although a right transverse colostomy has been advocated by some surgeons as the initial procedure, the authors prefer a leveling colostomy. This allows for the determination of the aganglionic level at the time of the colostomy, facilitating the subsequent pull-through. In addition, placement of a leveling colostomy allows the proximal bowel to grow, which will stretch the mesentery and simplify the subsequent pull-through procedure. Finally, this colostomy can be closed during the pull-through, thus avoiding a third operation. The ostomy should be created just proximal to the transition zone. Use of laparoscopy, with serial seromuscular biopsies, has greatly facilitated the determination of the level of aganglionosis (see **Video 47.1**). This allows the surgeon to place the colostomy in the most optimal site. If an open procedure is performed, an oblique left lower quadrant incision is preferred. If the level of aganglionosis is not readily apparent on rectosigmoid biopsy, this incision can be extended transversely across the midline to aid in exposure of more proximal colon.

Preoperative

Typically, the diagnosis of Hirschsprung disease can be suspected based on the history alone, but confirmation is required by histopathologic examination of a rectal biopsy. The infant should receive rectal washouts, and broad-spectrum IV antibiotics should be given just prior to the incision. No formal bowel preparation is required.

Operative technique

Upon exploration, an attempt should be made to define a gross transition zone. The bowel proximal to the transition zone is normally dilated and has a diffuse hypertrophy of the muscular layer, often with no clearly distinguishable teniae coli (**Figure 47.4**). In neonates, a transition may not be seen. If this is the case, a good starting point for biopsy is just above the peritoneal reflection.

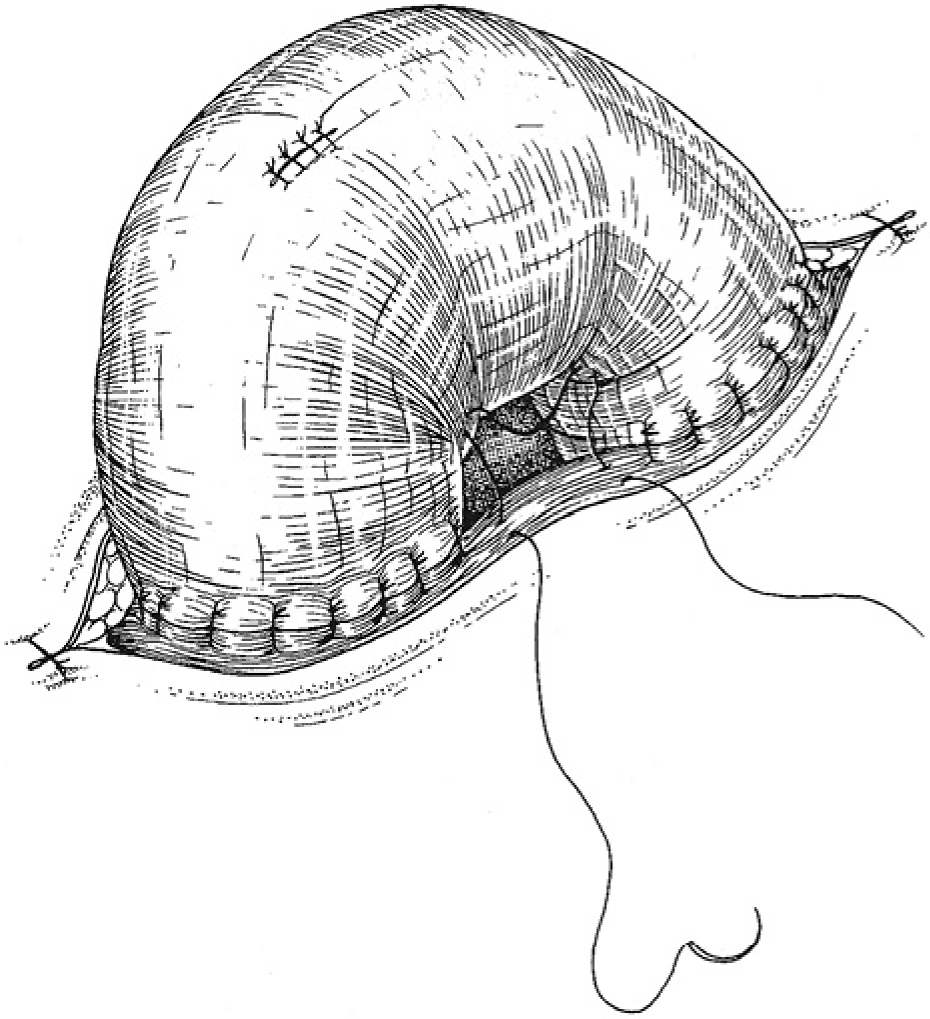


47.4

Whether laparoscopic or open, a pair of fine, sharp scissors is used to make an incision only through the seromuscular layers. The muscular layer is typically fairly thick even in the aganglionic section which makes this dissection fairly easy. In general, a 1 cm × 0.5 cm biopsy specimen is taken. Some advocate that closure is needed only for full-thickness biopsy. It is the authors' preference to close the biopsy site in all cases to prevent spillage but also to serve as a marker for later identification.

Each biopsy specimen is sent for frozen section, progressively moving more proximally until both ganglion cells, as well as a loss of hypertrophied nerve bundles, are seen. Hypertrophied nerve bundles, despite the presence of ganglia, indicate that one is still in the transition zone and another biopsy specimen should be taken several centimeters more proximally. Importantly, the transition zone varies from the antimesenteric and mesenteric sides of the bowel. Thus, the surgeon must confirm this correct level by sending a frozen section on the mesenteric side of the colon as well.

At this point, an end or loop colostomy can be created at the normal biopsy site. Because of the relatively large caliber of the bowel, stomal prolapse and peristomal hernias are common complications. It is extremely important to begin the colostomy by placing numerous fine polyglactin sutures both to the peritoneum as well as to the fascia. In the case of a loop colostomy, a stitch is placed between the proximal and distal loops of bowel, starting at the fascia, then to each limb of bowel, and finally back to the fascia (**Figure 47.5**). A portion of the fascia may be closed in the mid-portion of the defect between intestinal segments. If an end colostomy is preferred, a long Hartman's pouch can be left in place or aganglionic colon can be resected to just above the peritoneal reflection.



47.5

Postoperative care

The stoma usually begins to function within 24 hours, and feeding can begin shortly thereafter. It is occasionally useful to perform intermittent dilatations of the proximal ostomy. These dilatations will prevent narrowing of the opening and allow the dilated proximal colon to return to normal size.

Duhamel

The Duhamel technique was advanced in 1956 to avoid the tedious pelvic dissection of the Swenson procedure, and to protect the nervi erigenti, which may be found lateral and anterior to the rectum. The procedure has undergone several modifications, the most important of which was by Martin and included the use of an automatic stapling device. It is fairly straightforward and continues to be popular today. Despite its relative simplicity, several key technical points must be followed.

As with other pull-through procedures, ganglionic bowel is brought down to less than 1 cm proximal to the dentate line. To preserve the autonomic nerve plexus to the genitourinary system, very little manipulation of the rectum is performed anteriorly.

The operation is generally performed when the child is 6–12 months of age with a weight of 10 kg. With the use of smaller endostapling devices, the procedure may also be performed primarily in the newborn period.

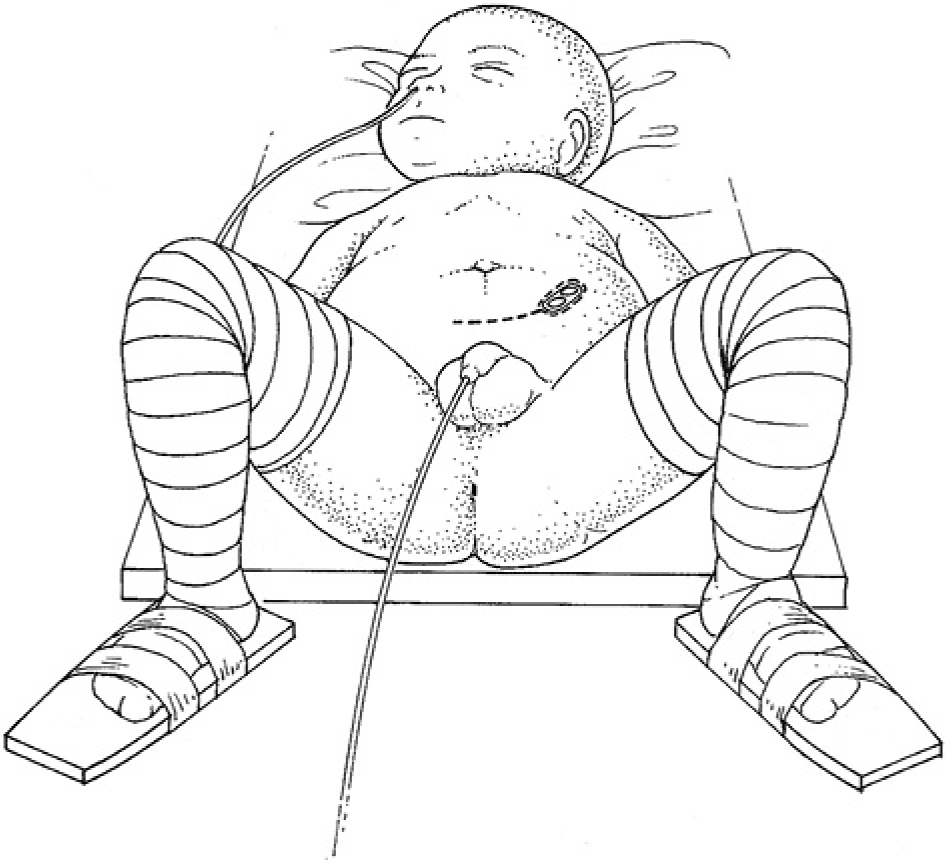
PREOPERATIVE

The child is admitted the day before the surgery for a mechanical bowel preparation as well as oral antibiotics. More recently, based on lack of evidence supporting its use, many have omitted the use of a bowel prep. Care must be taken to give adequate rectal and colonic washouts, as stool is often inspissated in the distal rectum. It is necessary to do a rectal examination on the child before the pull-through to ensure that no residual stool is present. Preoperative broad-spectrum antibiotics should be given just prior to the skin incision.

OPERATIVE TECHNIQUE

A nasogastric tube is placed after induction of anesthesia. The child is placed in a supine position and prepared circumferentially from the abdomen to the feet. Stockinettes are placed around each foot and a Foley catheter is inserted in the bladder after the patient has been prepared and draped (Figure 47.6). Excellent exposure is obtained by assistants supporting and flexing the lower extremities at the hips during the anal anastomosis. Alternatively, the child can be placed in stirrups or on skis.

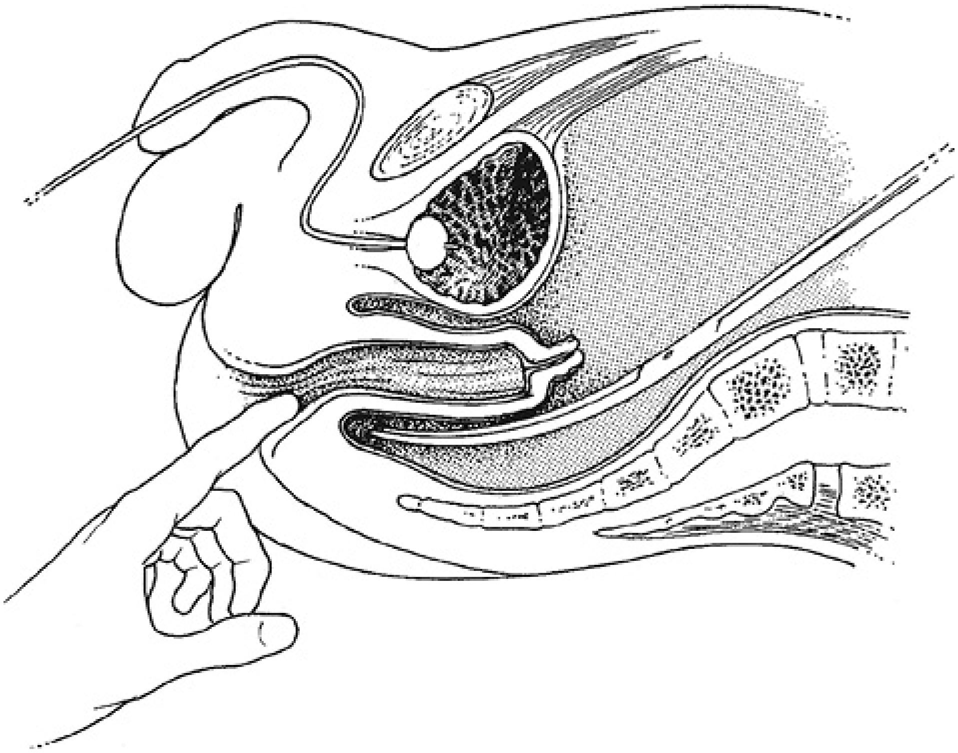
The circumferential prep allows for prone positioning for the perineal portion of the procedure, should the surgeon desire.



47.6

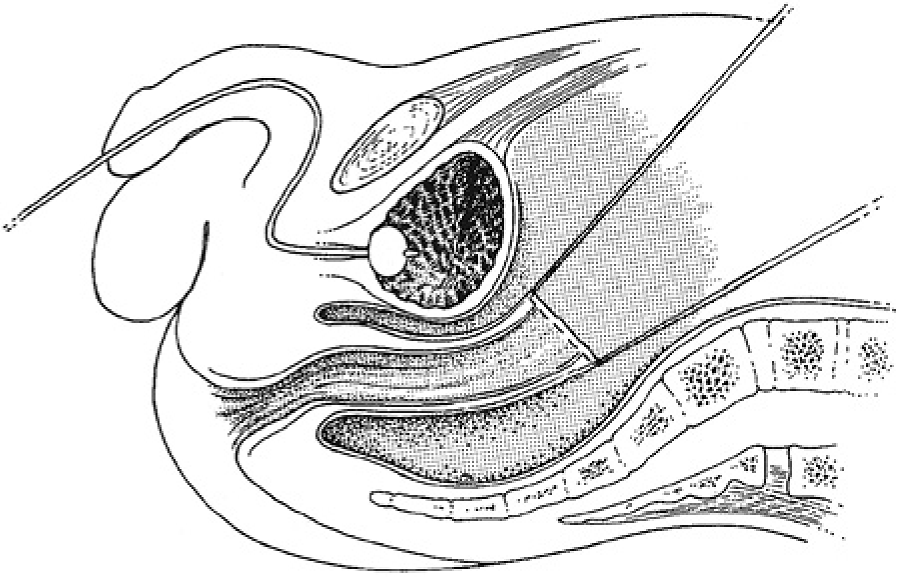
A hockey-stick or oblique incision is made incorporating the colostomy (if present). The bowel is mobilized proximal to the former colostomy and the splenic flexure is brought down, if necessary, to ensure adequate length for the pull-through. In general, the ganglionated colon must reach just below the level of the perineum when drawn over the child's pubic symphysis with only modest tension. Occasionally, the mesentery is foreshortened and it is necessary to ligate the inferior mesenteric artery near the aortic root. By preserving the remainder of the arcades, the bowel should maintain its viability. The ureter is carefully identified and the peritoneal reflection between the rectum and bladder is incised. The distal rectum is mobilized for approximately 4 cm below the reflection. The colostomy site, if present, is removed with an automatic stapling device.

A retrorectal space is created, with dissection carried out directly in the midline. This dissection is carried down to the pelvic floor so that an assistant's finger can be felt when inserted no further than 1–1.5 cm into the anus (**Figure 47.7**). Dissection can be facilitated by a blunt clamp, but is also very easily performed with the index finger.



47.7

Once the retrorectal dissection has been completed, redundant aganglionic bowel is resected down to the peritoneal reflection with an automatic stapling device. Tacking sutures are placed on both left and right sides of the remaining distal bowel so that it can be retracted anteriorly during the pull-through procedure (**Figure 47.8**).

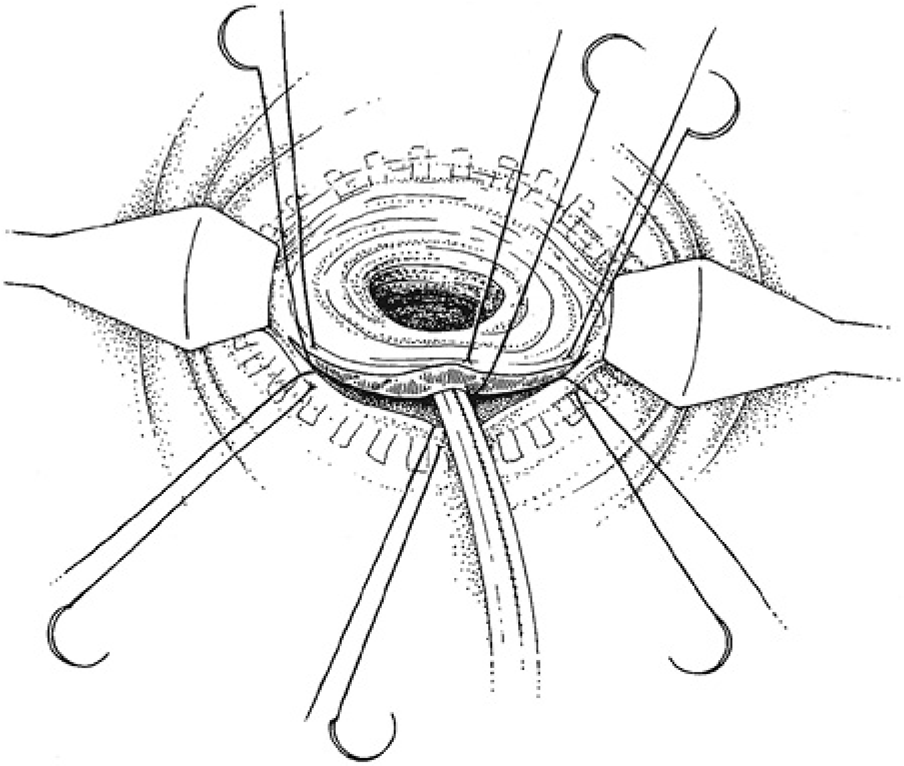


47.8

The distal end of the ganglionic bowel is labeled mesenteric and antimesenteric with a separate polypropylene and polyglactin suture. This allows the surgeon working on the pulled-through segment to maintain correct orientation of the bowel as it is pulled into the anus.

At this point, the surgeon's attention is directed to the perineum. If not placed into skis, both the patient's legs are drawn upward, allowing a clear view of the anus. Narrow anal retractors are placed and held in position by the two assistants still working on the abdomen. Care should be taken not to overstretch the anal opening, which could result in damage to the sphincter complex. No separate field is created, which allows improved communication and keeps surgeon and assistant on the same operative field. The authors have not found it necessary to have two completely different set-ups. However, all the instruments used in the perineal portion of the procedure are treated as dirty, and gloves are changed at the end of the anastomosis.

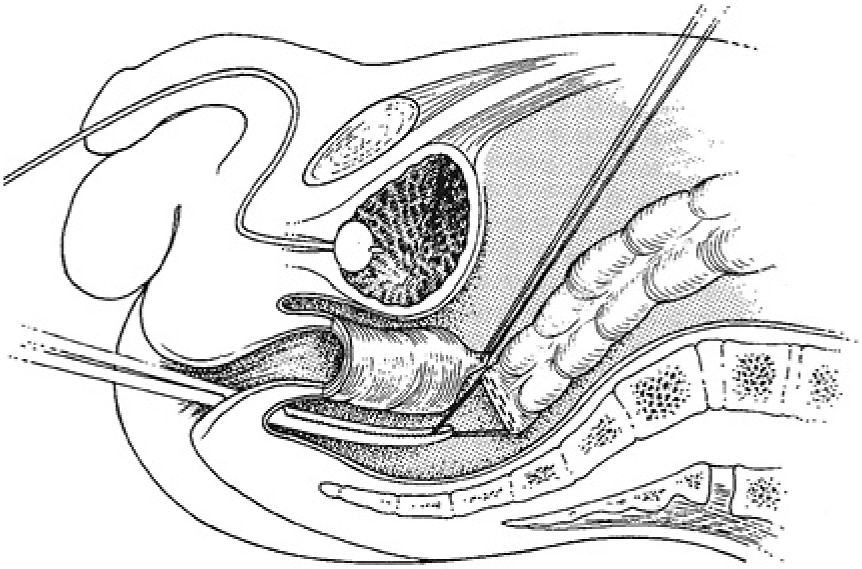
With the use of cautery, a full-thickness incision is made 0.5 cm proximal to the dentate line posteriorly. Care is taken to maintain this distance by curving the incision as one moves laterally in each direction. Three 4/0 undyed polyglactin sutures are placed on the inferior aspect of this incision, one in the midline and one each on the left and right sides (**Figure 47.9**). Three additional absorbable sutures (4/0 dyed polyglactin) are placed on the upper portion of this incision in similar positions. Needles are placed into the defect created by the incision and are retained for the initiation of the anastomosis. Each suture is held in position with hemostats. The different-colored sutures prevent confusion of orientation once the ganglionic bowel is pulled through.



47.9

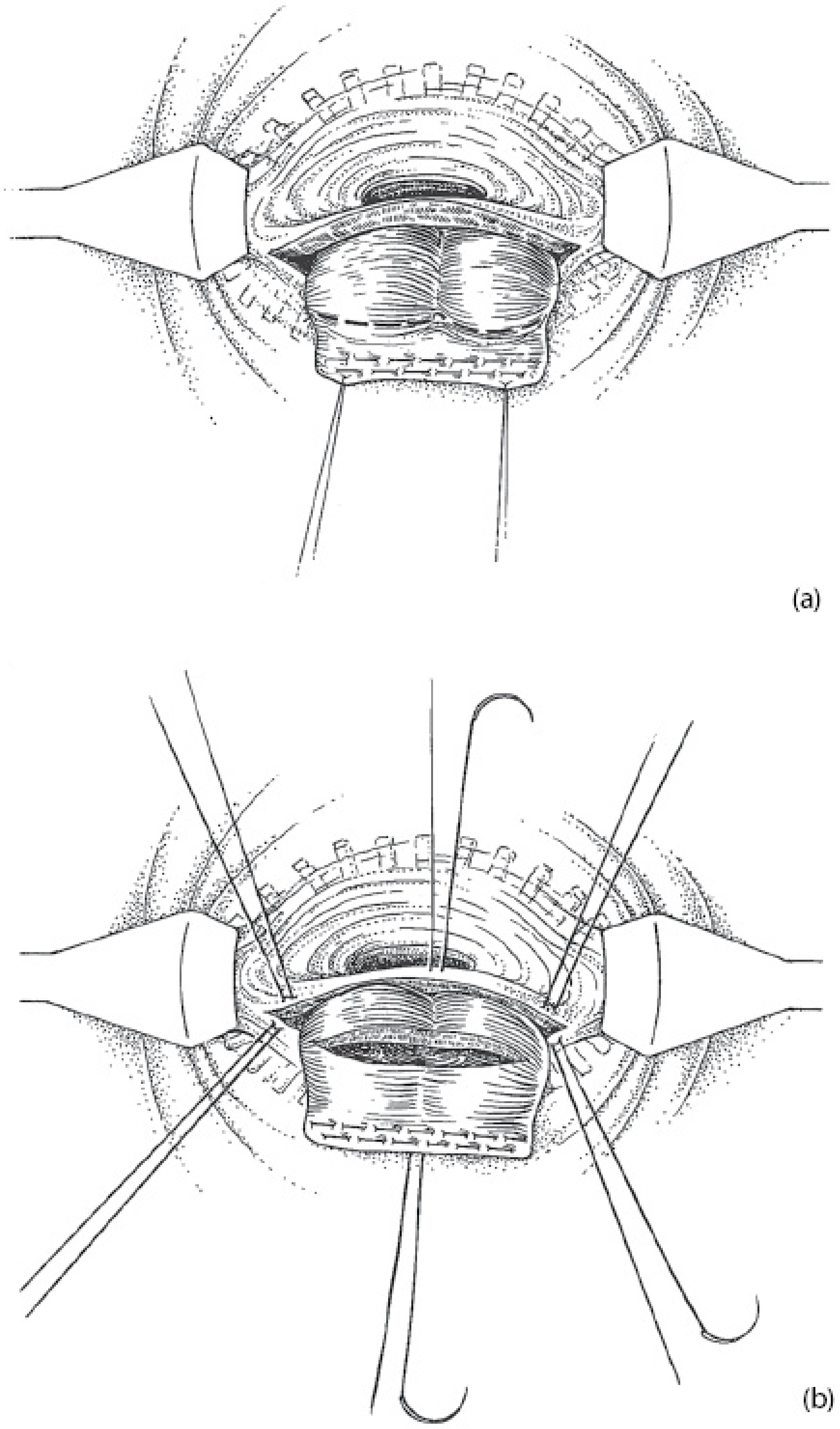
The surgeon operating on the anus inserts a long clamp into the retrorectal space toward the abdominal field.

The two tacking sutures on the distal ganglionic bowel are fed into this clamp and pulled down (**Figure 47.10**). The surgeon remaining in the abdominal field guides the bowel, and makes sure that it does not rotate as it is brought down.



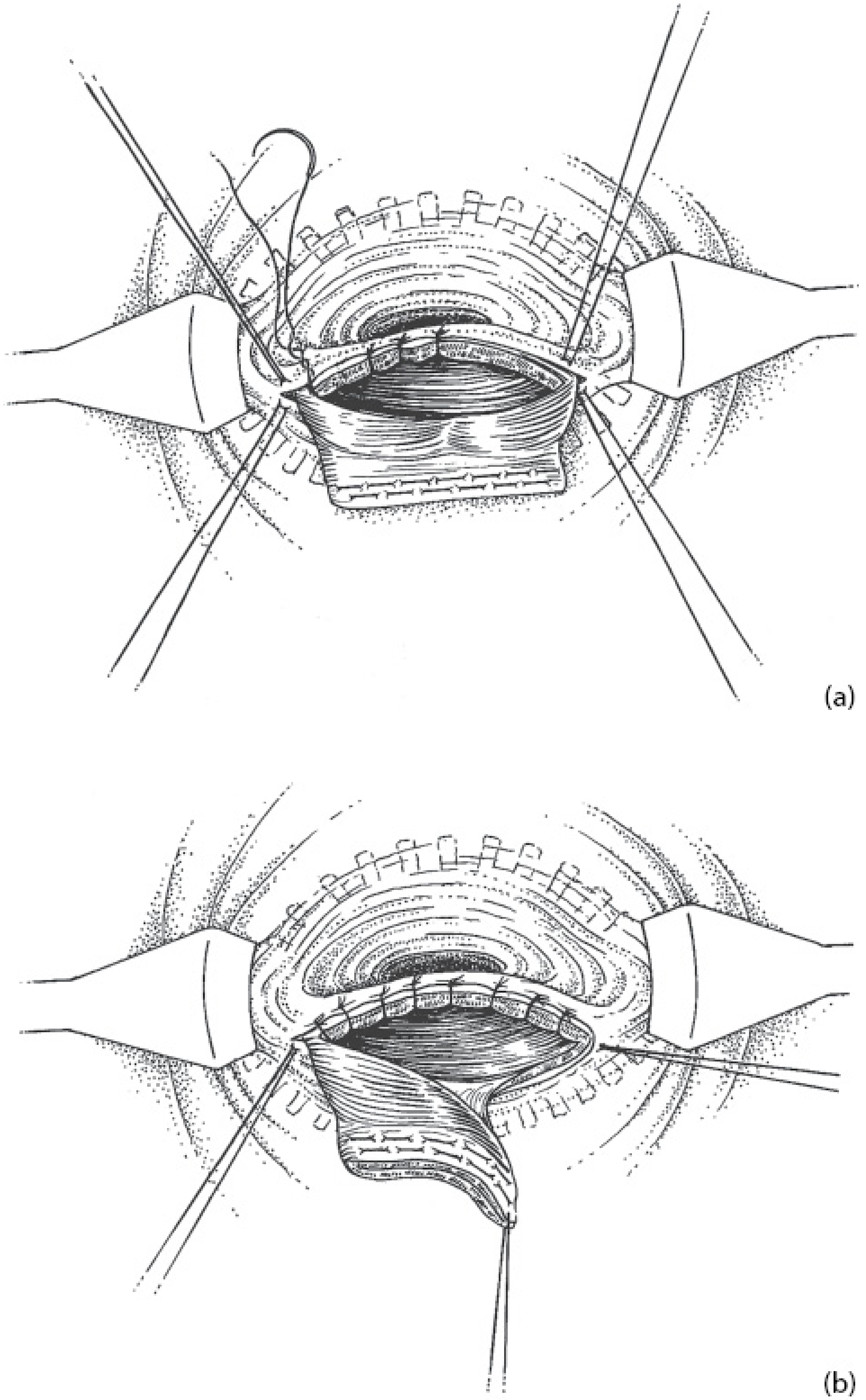
47.10

Once the bowel is pulled through, the staple line is excised on the anterior half of the colon and a single-layered anastomosis is created, starting with the three previously placed polyglactin sutures (**Figure 47.11a and b**). Care is taken with each stitch so that the anterior wall of the anus is not incorporated into any of the sutures.



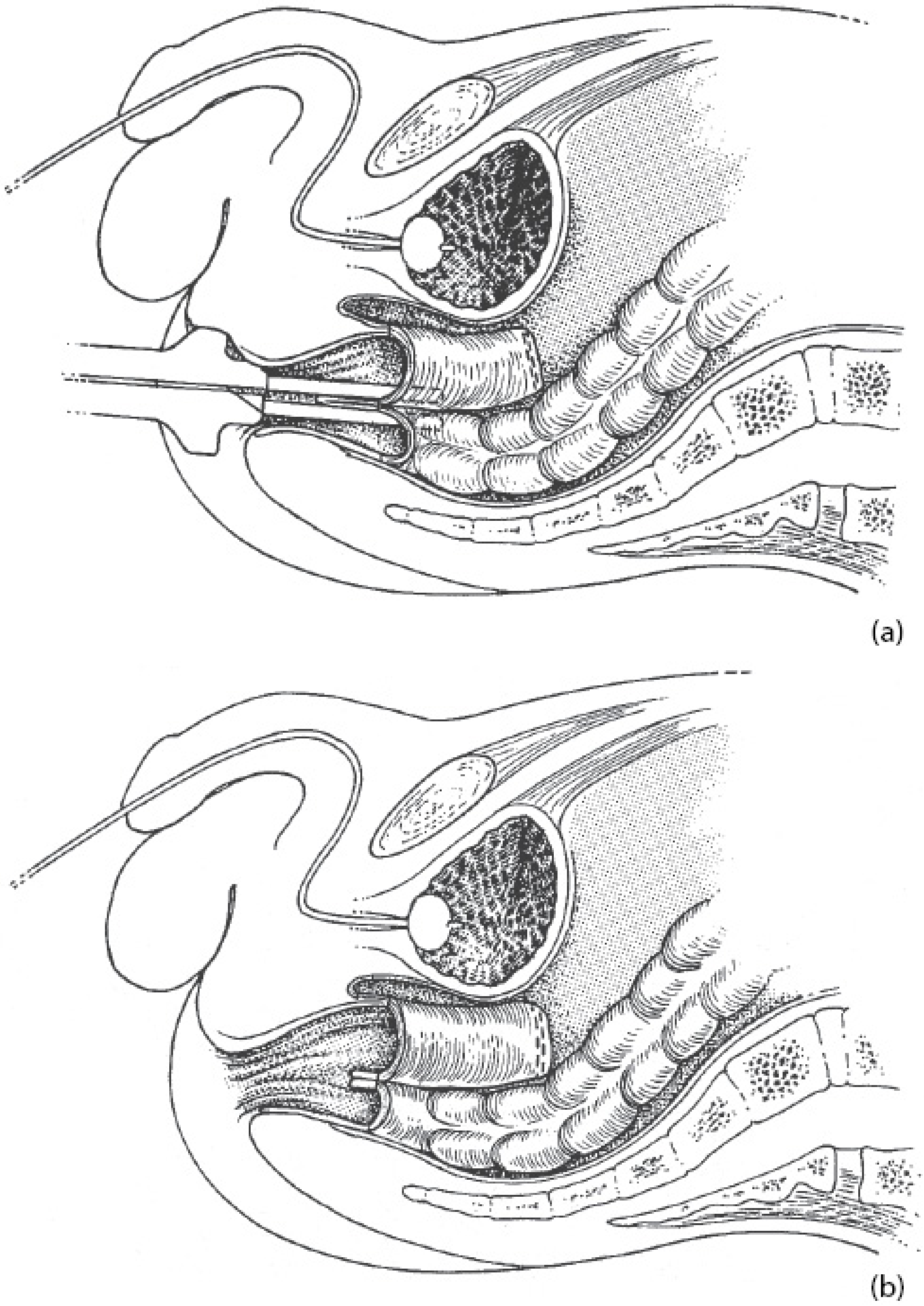
47.11a,b

Once the anterior half of the anastomosis is completed, the remainder of the staple line is excised, one-quarter at a time, and the anastomosis is finished (**Figure 47.12a and b**).



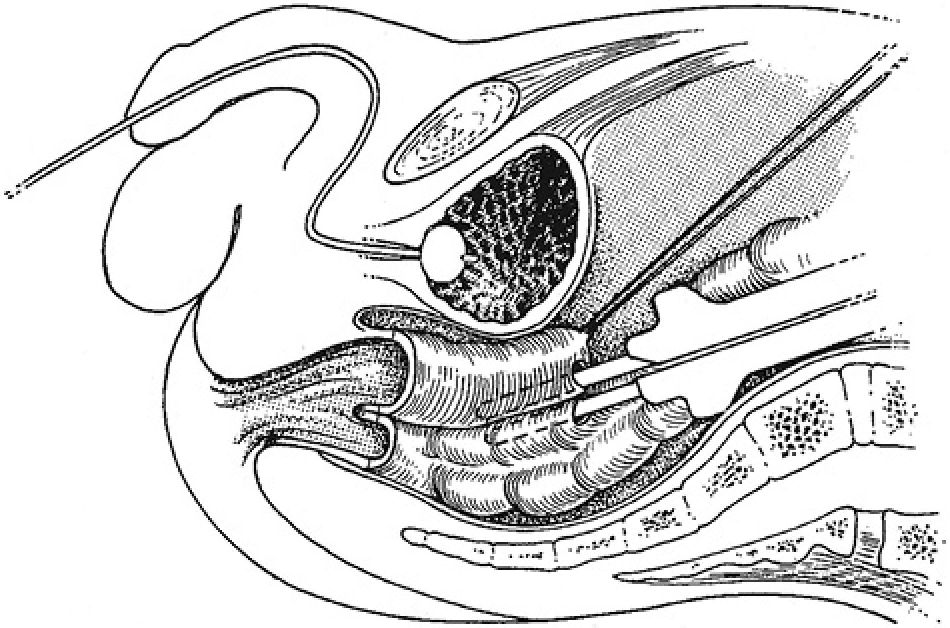
47.12a,b

An automatic stapling device is placed with one arm into the native anal canal and the other into the neorectum along the antimesenteric aspect (**Figure 47.13a**). The stapler is fired directly in the midline (**Figure 47.13b**). Hemostasis along the suture line is checked. A long (80 mm with 3.5 mm staples) device is generally preferred; a smaller endostapler is used in newborns.



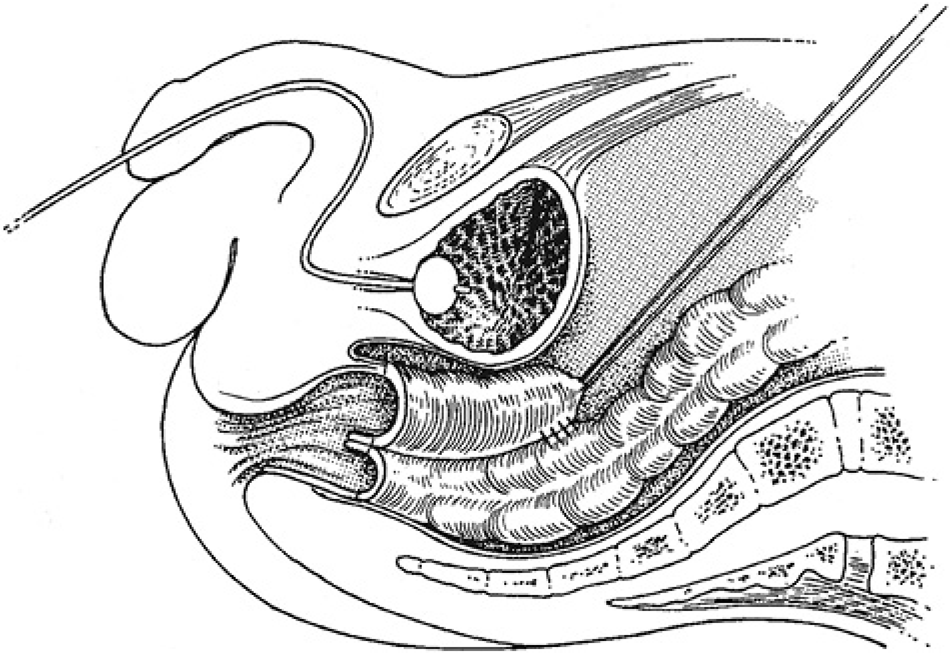
47.13a,b

Often a complete anastomosis between the ganglionic and aganglionic bowel cannot be achieved with a single staple application from below. A second firing from the abdomen may be necessary. The staple line of the remaining aganglionic rectum is opened and a small enterotomy is made in the ganglionic colon at a similar level. The abdominal surgeon places a reloaded automatic stapler between the two limbs of bowel to complete the anastomosis (**Figure 47.14**). This last step is critical. In the past, a proximal spur left between the bowel segments caused the eventual formation of huge fecalomas. It is critical for the surgeon to inspect the anastomosis digitally and make sure that the two limbs of bowel are completely anastomosed.



47.14

The anastomosis is completed by suturing the proximal end of the aganglionic rectum to the enterotomy in the ganglionic colon in two layers (**Figure 47.15**). The neorectum may or may not be reperitonealized, and the abdomen is closed.



47.15

Endorectal pull-through (Soave)

The Soave or endorectal pull-through was popularized by Franco Soave at the Institute G. Gaslini in 1955. The procedure was modified by Boley by performing a primary anastomosis at the anus, and then further modified by Coran. This procedure is the most popular one used at the authors' institute, and has been further modified to facilitate the suturing of the anal anastomosis. The operation is now most commonly performed in the newborn period, with the complication rate being identical to that seen with the standard two-staged approach. Over the past two decades, the operation has been performed with laparoscopic assistance and via a transanal route.

As with the Duhamel technique, this procedure avoids injury to the pelvic nerves and, by remaining within the muscular wall of the aganglionic segment, important sensory fibers and the integrity of the internal sphincter are preserved. Although one imagines that leaving aganglionic muscle surrounding normal bowel could lead to a high incidence of constipation, this is generally not the case. Overtime, the length of the submucosal dissection and thus the muscular cuff has been shortened to avoid the postsurgical complications of an obstructing cuff. Most surgeons performing this technique routinely leave a 0.5–3 cm Soave cuff with most leaning toward the former. Additionally, most split or resect a portion of the posterior aspect of the cuff.

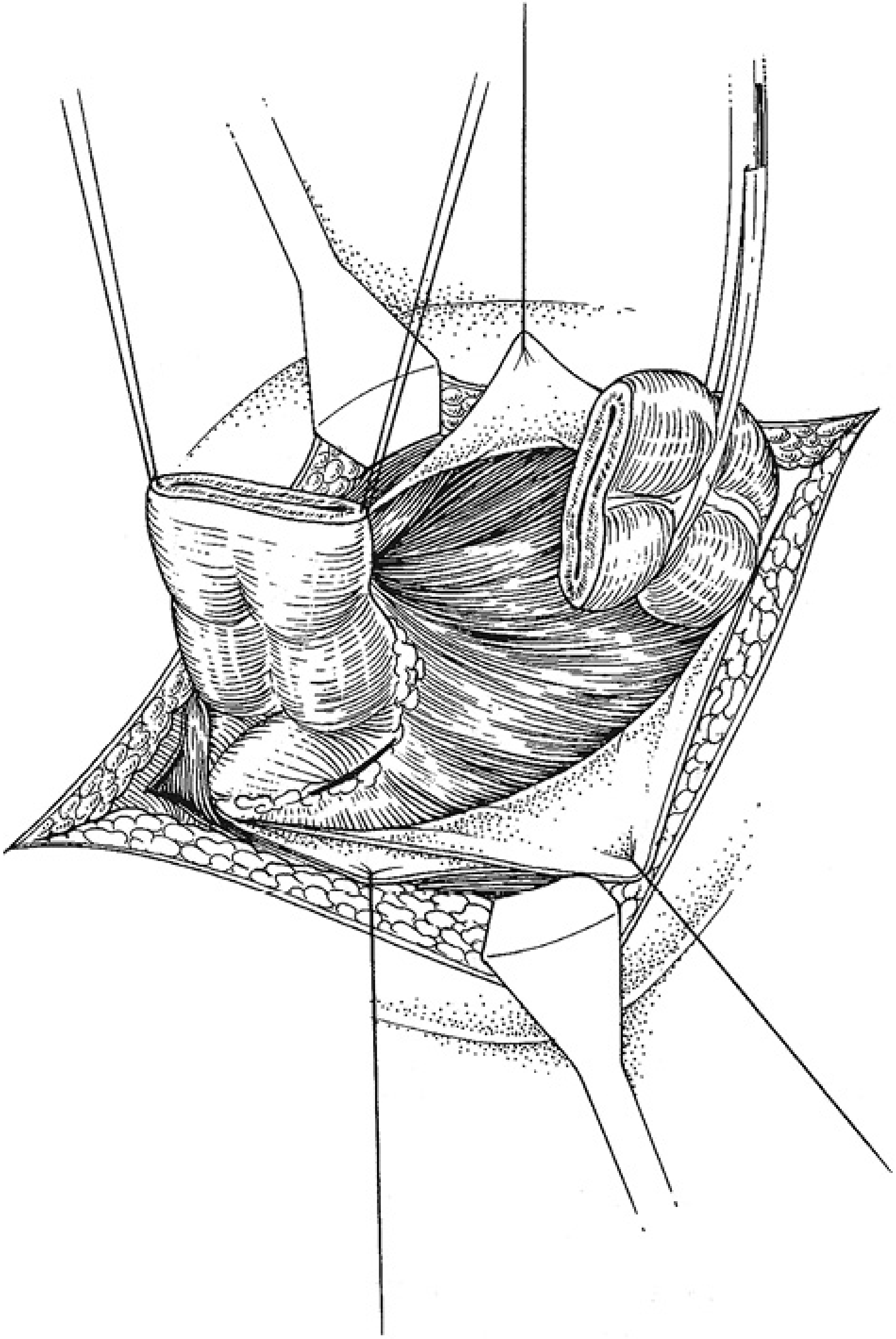
PREOPERATIVE

Even in the neonatal period, serial rectal washouts and digital dilatations of the rectum are performed before beginning the pull-through. The last of the rectal irrigations may or may not contain 1% neomycin. Intravenous antibiotics are given before the beginning of surgery and are continued for two doses after surgery.

OPEN OPERATIVE TECHNIQUE

The child is placed in a supine position, with the buttocks brought to the end of the operating table, and propped slightly up with a folded towel. The legs are carefully padded and placed on wooden skis extending off the end of the table. A Foley catheter is placed after the entire field is prepared and draped. The operating table is placed in a slightly Trendelenberg position. A hockey-stick incision is made incorporating the leveling colostomy (if present). The same type of incision is made, however, for infants undergoing a primary pull-through operation. The level of aganglionosis is established by frozen section.

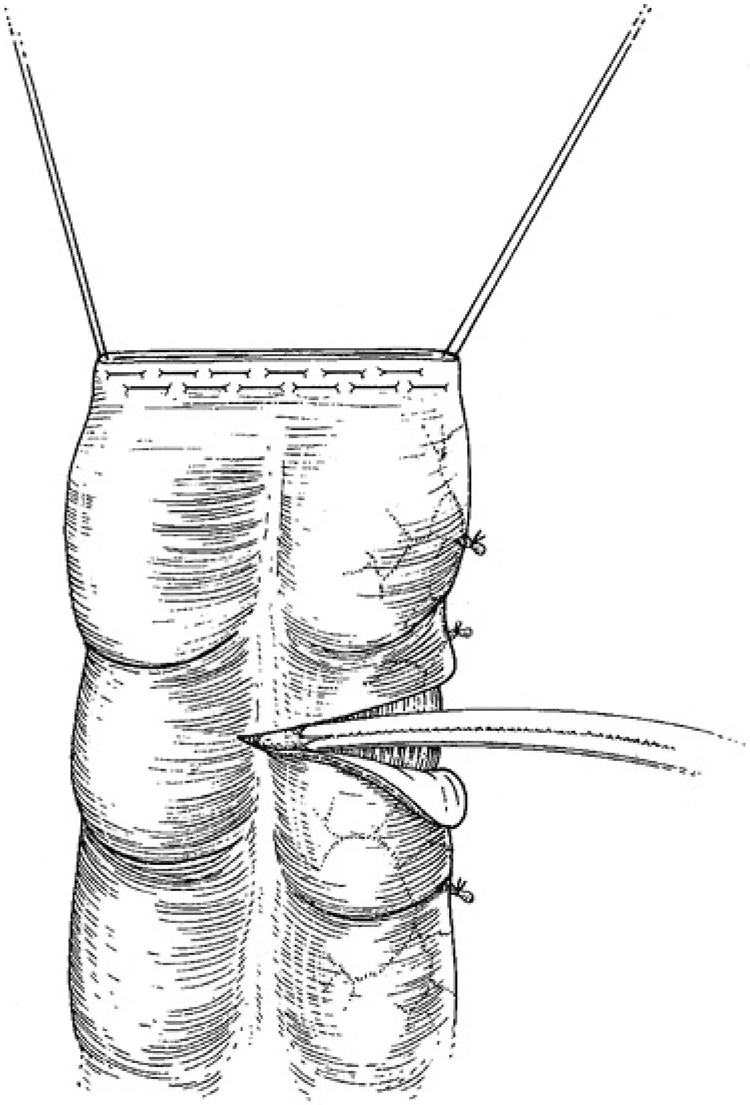
Ganglionic bowel is mobilized proximally and then transected above the transition level with a stapling device. The distal colon is mobilized and resected to about 4 cm above the peritoneal reflection. Traction sutures are placed on either end of the distal bowel. The endorectal dissection is then started about 2 cm below the peritoneal reflection (**Figure 47.16**).



47.16

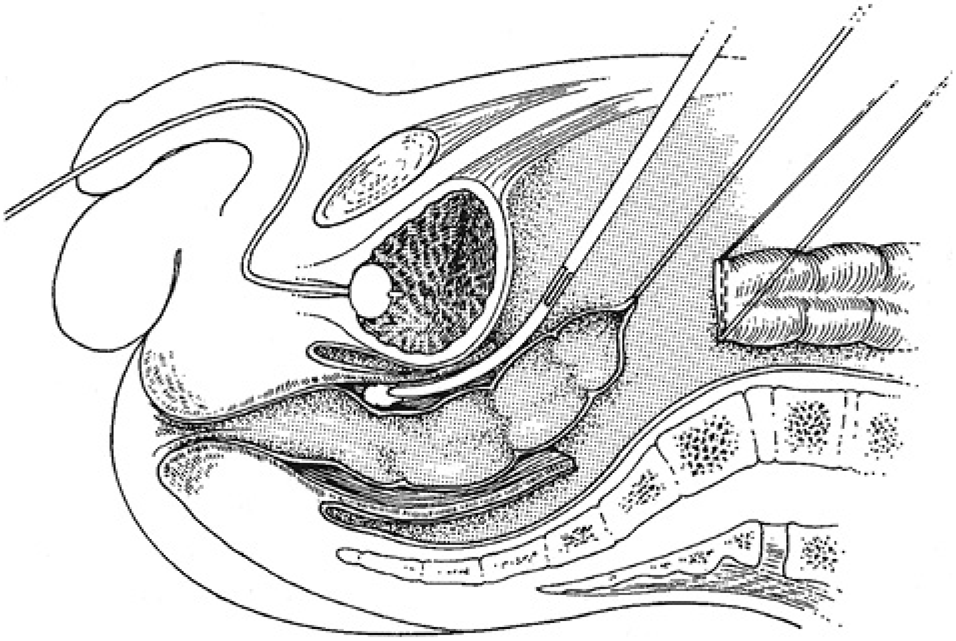
As stated above, the authors have progressively shortened the length of the endorectal dissection, because longer lengths of muscular cuff may lead to increased bouts of constipation and enterocolitis due to an obstructing cuff.

The endorectal dissection usually begins by completely clearing the serosa, mesentery, and fat over a 2 cm length of bowel. The seromuscular layer is incised with either sharp dissection or Bovie cautery (**Figure 47.17**). Once the submucosal layer is reached, the seromuscular layer is divided circumferentially using blunt dissection with hemostat or a Kitner dissector. In the neonatal period, a cotton-tip applicator is the most effective tool for this dissection.



47.17

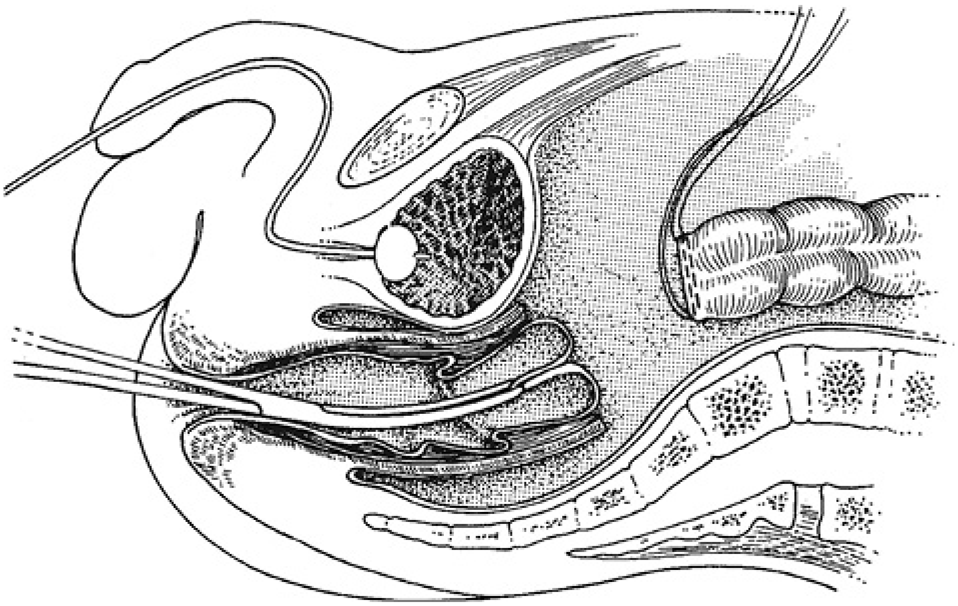
After the plane is established, it is continued distally and facilitated by an assistant pulling upward on the already dissected mucosal/submucosal tube for countertraction (**Figure 47.18**). As the muscular cuff begins to develop, traction sutures are also placed in the muscle, one in each quadrant. Larger communicating vessels are coagulated; however, the majority of these are not cauterized during the dissection without resultant significant blood loss, particularly in the neonatal period. Dissection is carried down to within 1.5 cm of the anal opening in older children and approximately 1 cm in neonates.



47.18

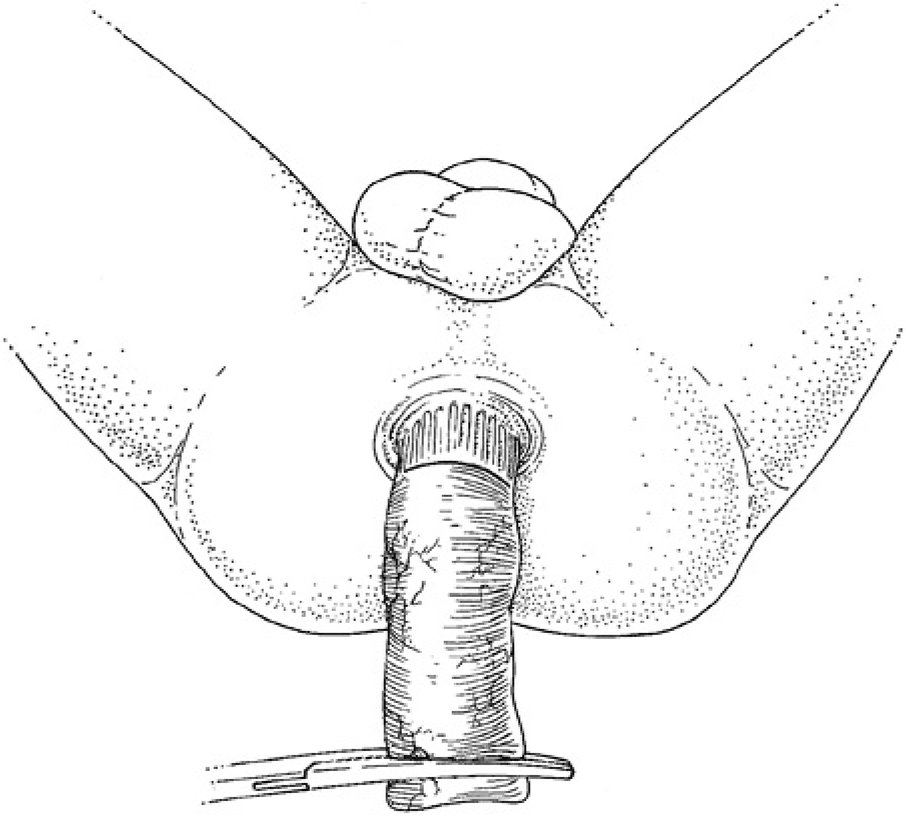
Some have advocated performing part of the endorectal dissection from the transanal approach, but the authors strongly advise against this if the dissection is started within the abdominal cavity. Once the endorectal dissection from above is started, it can proceed in a straightforward fashion. With appropriate traction and countertraction, the entire dissection can be performed in a child (and adult) of almost any size.

One of the surgeons then moves to the foot of the table. Narrow retractors (phrenic or army–navy) are placed at the anal mucocutaneous junction and a ring or Kelly clamp is inserted into the rectum (**Figure 47.19**). An assistant at the abdominal field places the end of the mucosal/submucosal tube into the clamp. The segment is then everted onto the perineum. The end of the everted tube is placed in a clamp and held on traction by an assistant to facilitate the anastomosis.



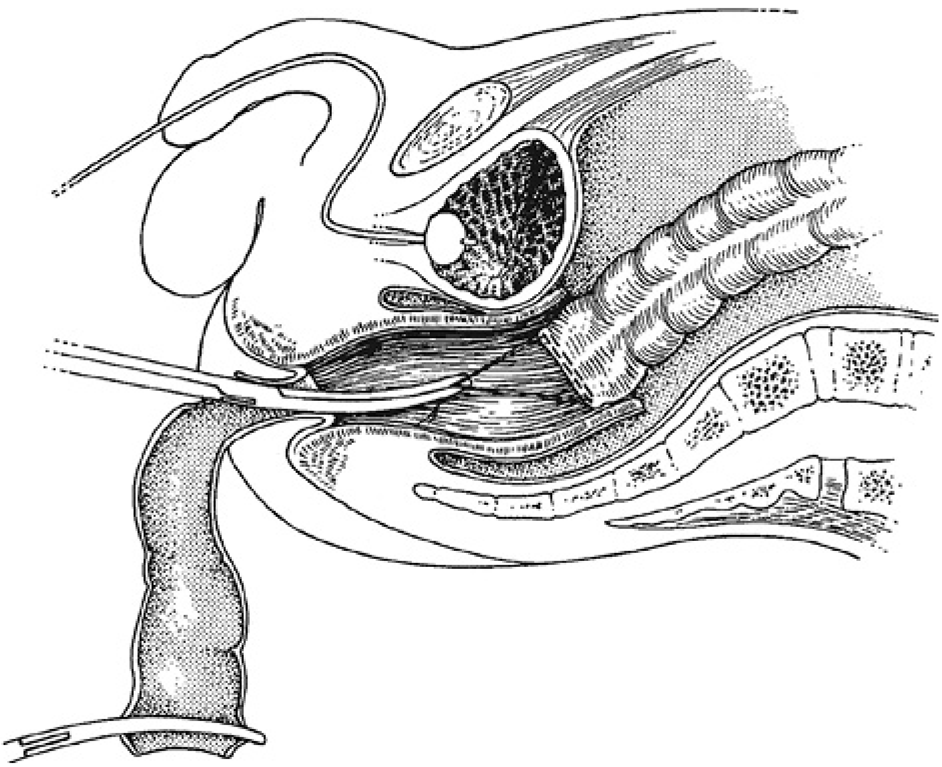
47.19

The mucosal/submucosal tube is incised on the anterior half, 0.5–1 cm above the dentate line (**Figure 47.20**).



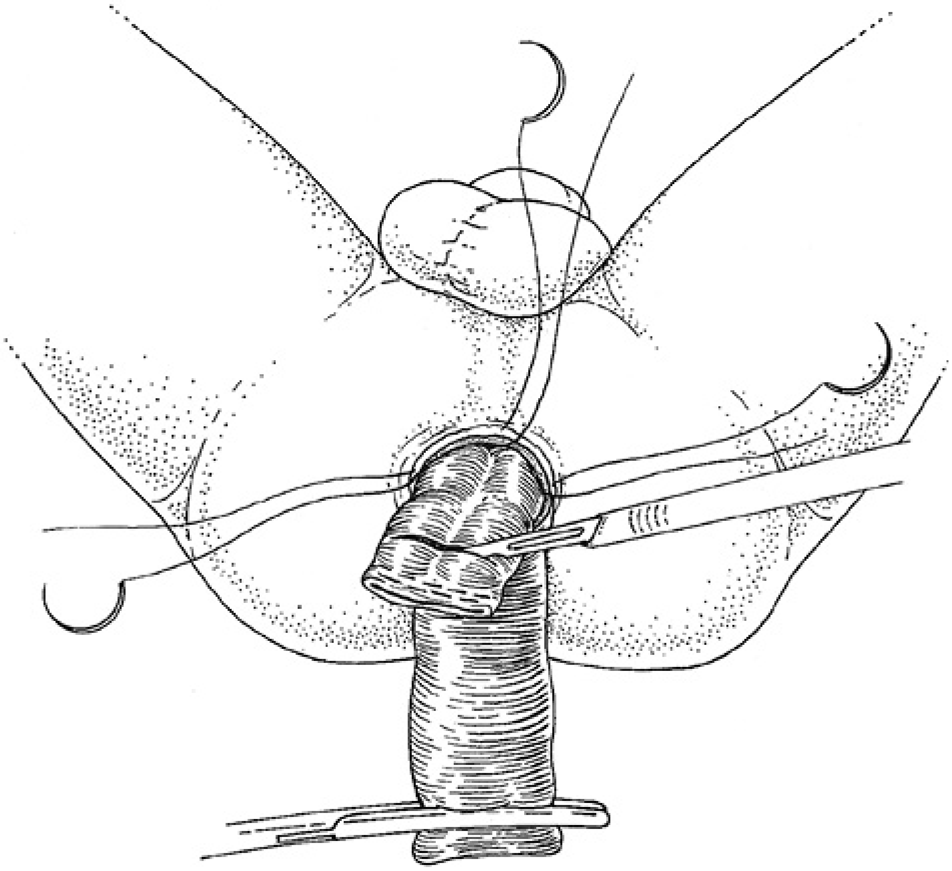
47.20

A Kelly clamp is inserted into this opening and the ganglionic bowel is brought down to this point by grasping the two previously placed traction sutures (**Figure 47.21**). Great care is taken not to twist the bowel as it is brought through the muscular cuff. As with the Duhamel procedure, different-colored sutures on each side of the bowel are helpful in maintaining orientation.



47.21

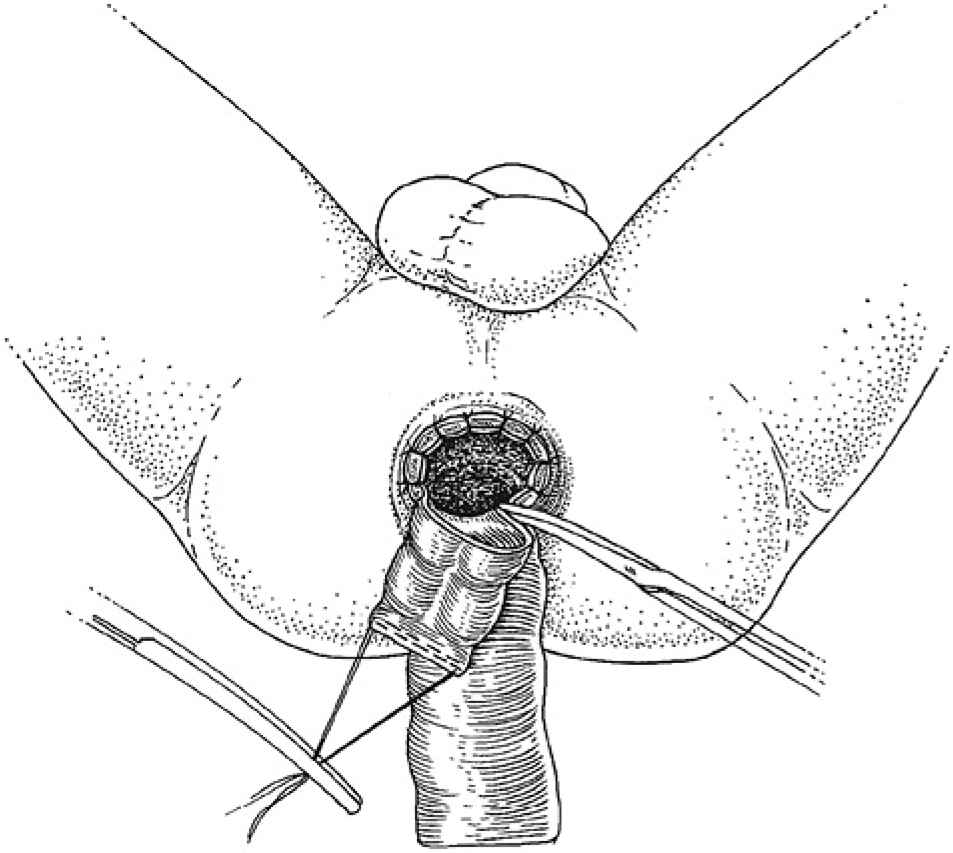
The anterior half of the ganglionic colon is incised and anastomosed to the anterior half of the anus with a 4/0 polyglactin suture (**Figure 47.22**). The first sutures are placed at each corner and in the midline, followed by interrupted sutures in between.



47.22

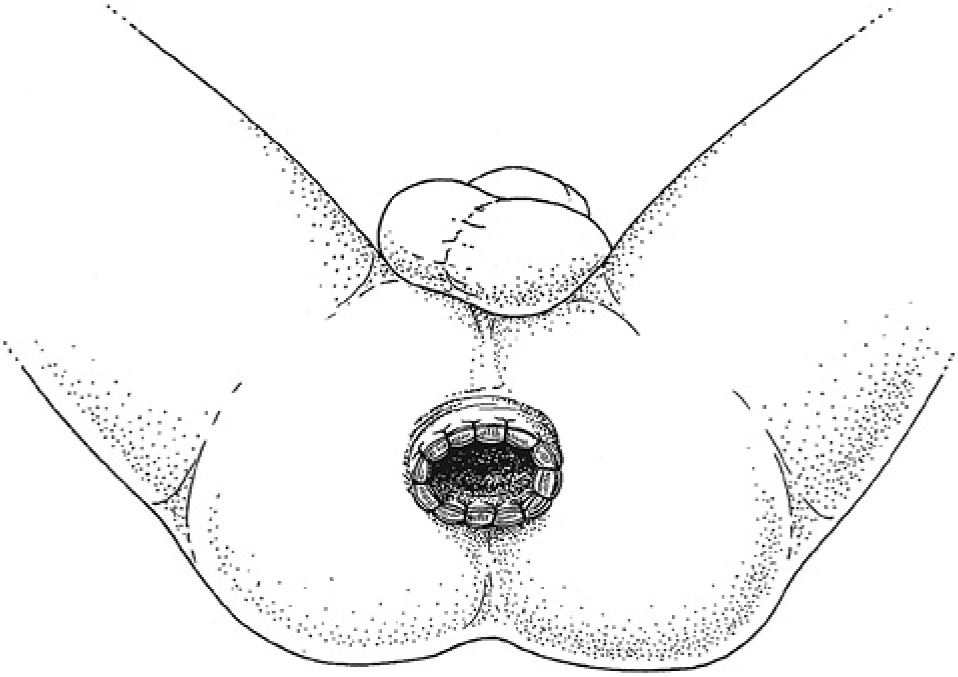
The assisting surgeon can facilitate visualization of the two edges of the bowel by putting outward traction on these initial quadrant sutures as the operating surgeon completes each quadrant.

One-quarter of the remaining ganglionic colon and one-quarter of the everted mucosal/submucosal tube are opened. A suture is placed in the posterior midline and this quarter of the anastomosis is completed (**Figure 47.23**). Throughout the anastomosis, countertraction applied by an assistant on the everted tube will help with the exposure.



47.23

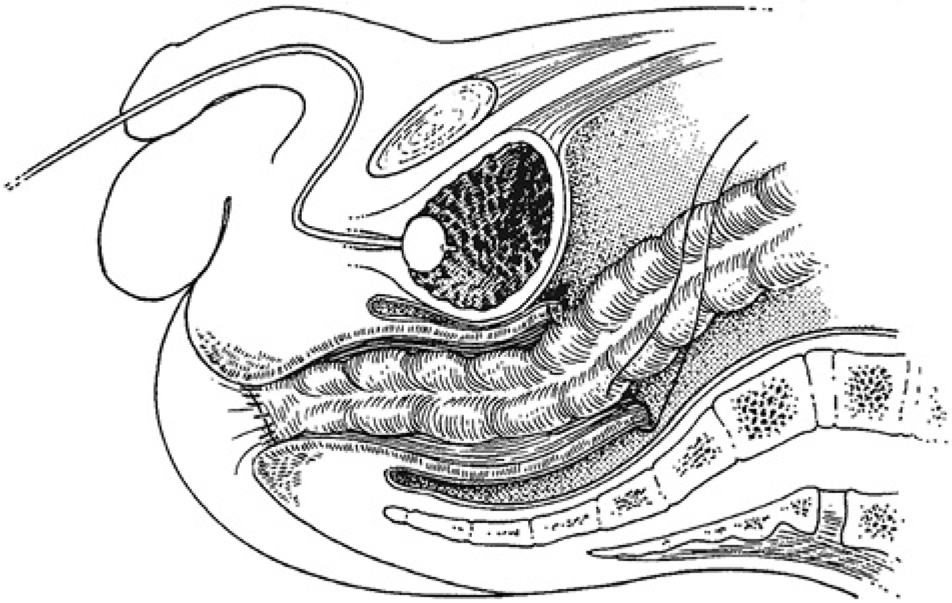
The final quadrant of the colon and submucosal/mucosal tube are removed, and the anastomosis is completed and inspected (**Figure 47.24**).



47.24

The colon is pulled slightly upward to invert the neorectum back into its correct position. Rectal examination at this point should reveal a well-formed anastomosis 1.5–2 cm above the anodermal junction. Gloves are then changed and attention directed to the abdominal field.

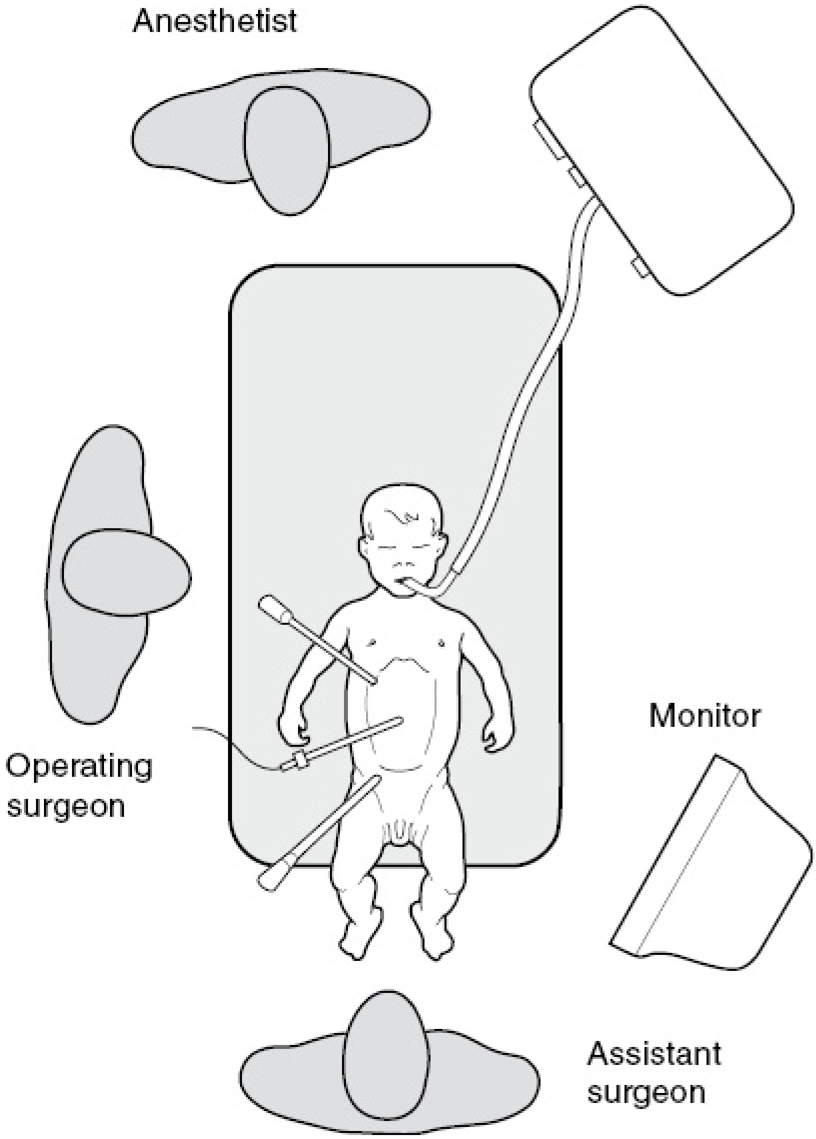
The pulled-through colon is attached with seromuscular bites to the muscular cuff to prevent the colon from prolapsing in the early postoperative period (**Figure 47.25**). In the cases of a shorter Soave cuff, similar stitches can be placed transanally in four quadrants. No drain is placed, either through the anastomosis or from above in the muscular cuff, because there is rarely any significant oozing in the cuff (unlike the situation with ulcerative colitis in older patients). Should the surgeon perform a traditional long-segment Soave, the muscular cuff must be split in the posterior position as distally as possible, generally within 1–2 cm of the internal anal sphincter.



47.25

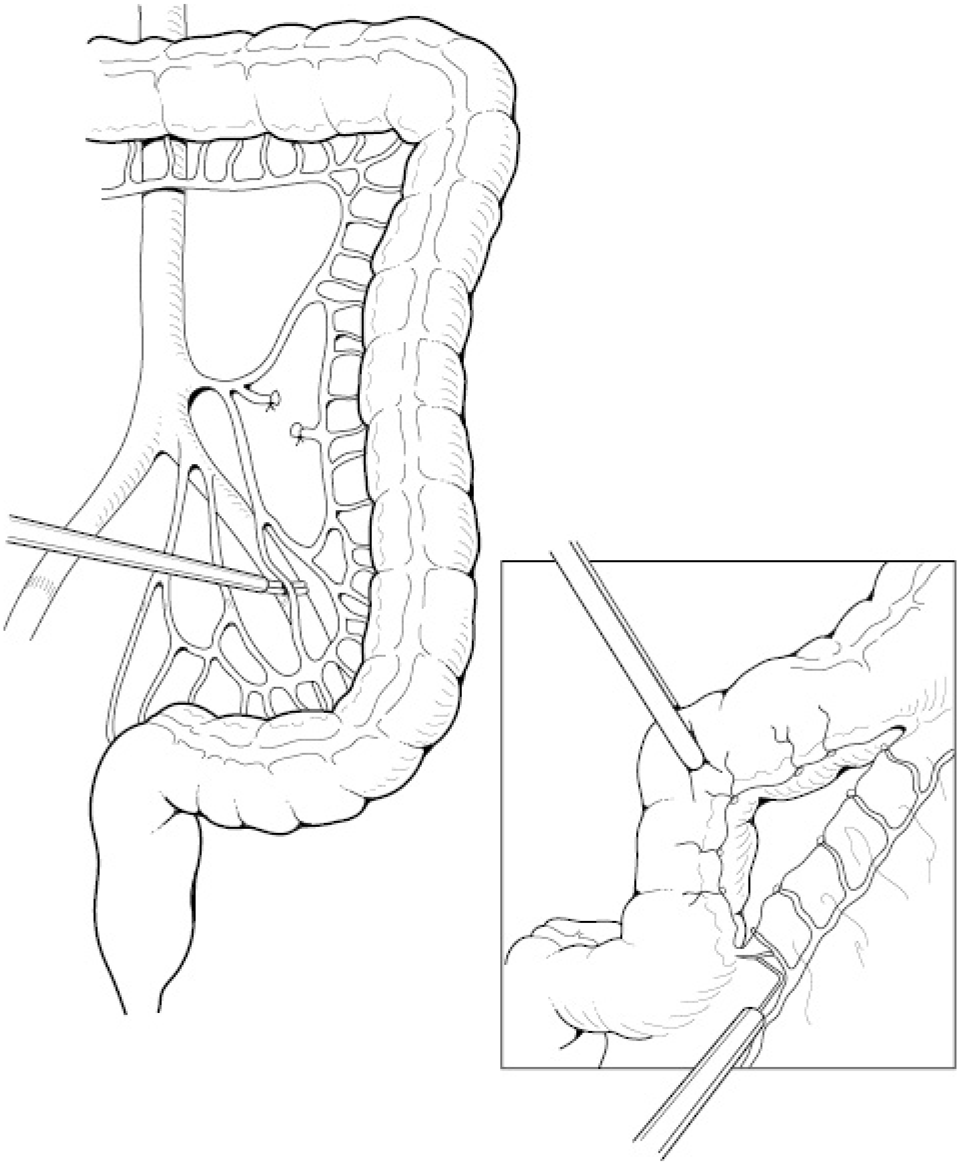
LAPAROSCOPIC APPROACH

The basic operative principles are virtually the same with the laparoscopic approach. Trocar placement consists of an initial umbilical trocar, followed by trocars in the right upper quadrant, and right lateral abdomen (**Figure 47.26**). An additional port can be placed in the left abdomen or in the suprapubic area to aid in retraction, but this is often unnecessary.


Anesthetist
Operating surgeon
Monitor
Assistant surgeon

47.26

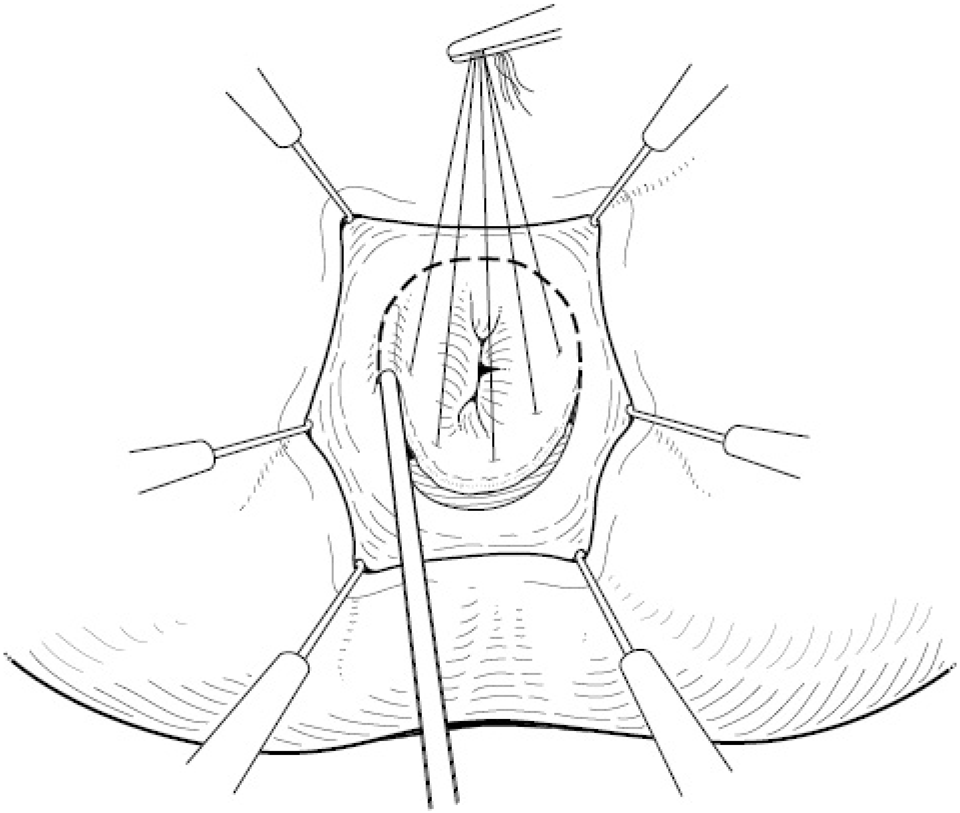
Dissection and leveling of the aganglionic segment are identical to the open technique. Blood vessels may be ligated with surgical clip appliers, or cautery for smaller vessels (**Figure 47.27**) (Video 47.1: Laparoscopic mobilization). Once mobilization is completed, the surgeon moves to the perineum, where a transanal dissection and anastomosis are performed (see "Transanal approach").



47.27

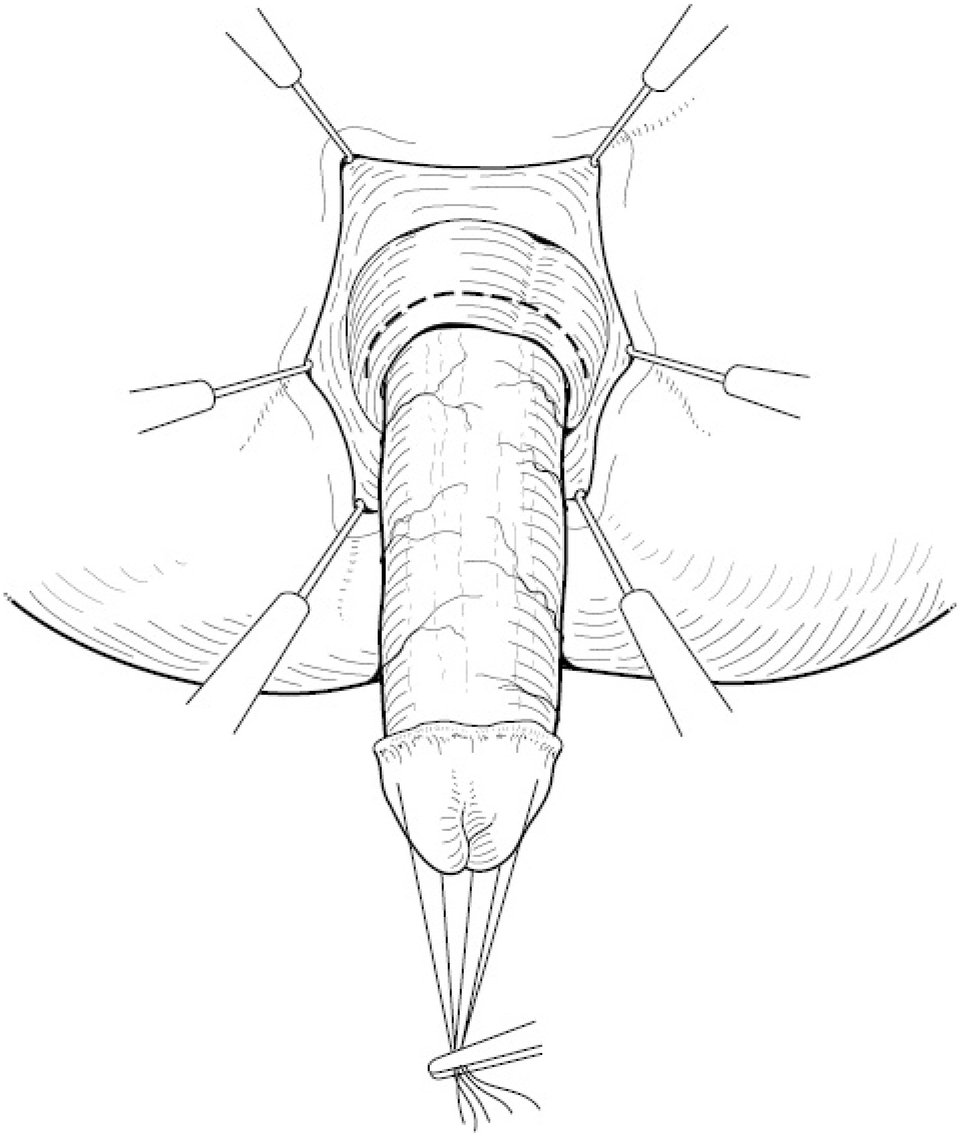
TRANSANAL APPROACH

The key element to beginning the case is placement of either a self-retaining retractor (e.g. Lone Star Medical Products, Houston, TX, USA) or a series of sutures that retract the anal verge (**Figure 47.28**). Caution must be taken if the decision has been taken to perform a transanal approach without laparoscopic leveling biopsy. Rarely, the surgeon can encounter long-segment or total colonic Hirschsprung disease. In some cases, an ostomy may be the best option in the neonatal period. However, if the operation is begun at the perineum, the surgeon has now committed to an anal anastomosis.



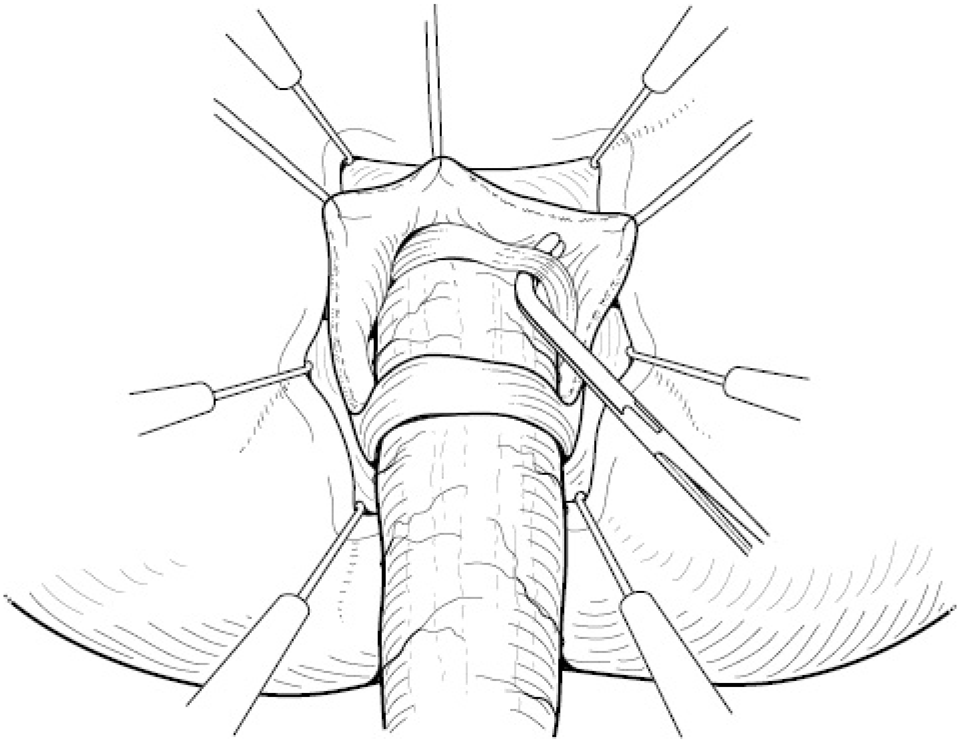
47.28

An incision 0.5–1 cm above the dentate line is begun with cautery in a circumferential fashion. A series of traction sutures are placed on the mucosa/submucosal tube, and dissection is carried proximally, primarily with blunt technique (**Figure 47.29**).



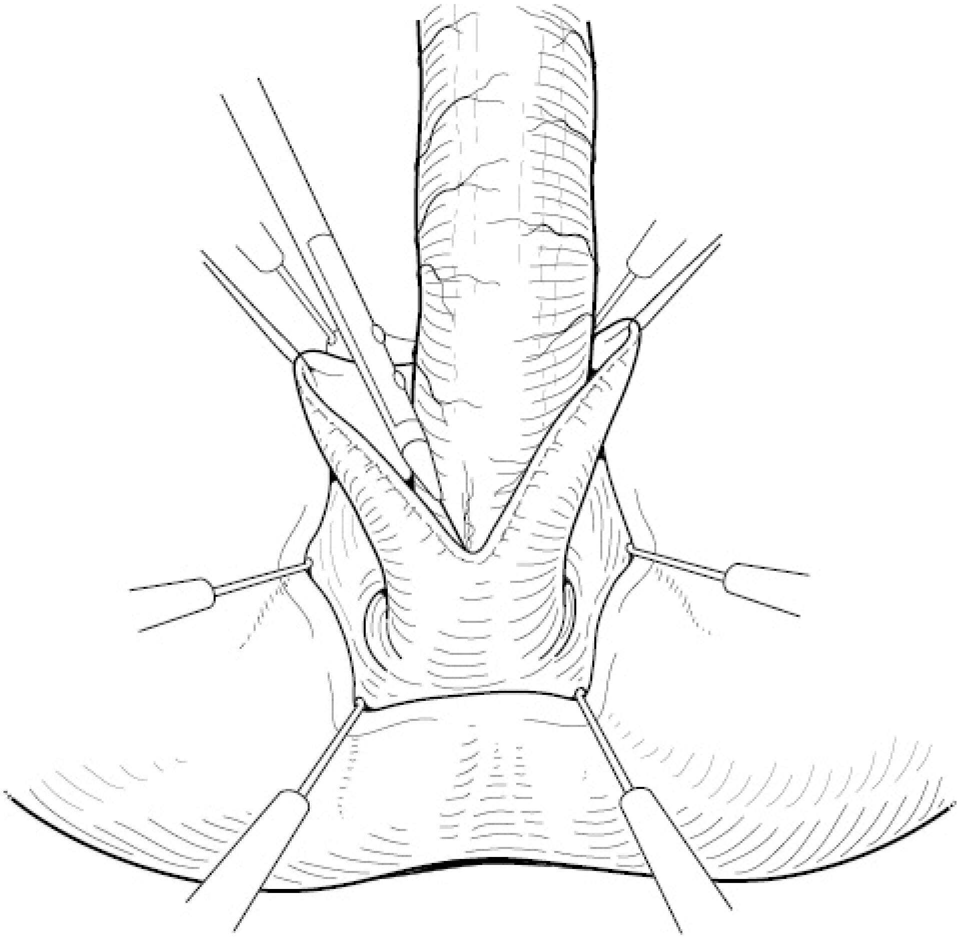
47.29

Once the dissection is carried proximally to the surgeon's preferred Soave cuff length, the muscular layer is entered posteriorly, and the dissection becomes full-thickness (**Figure 47.30**). This can be facilitated by placement of traction sutures on the muscular layer prior to dividing it.



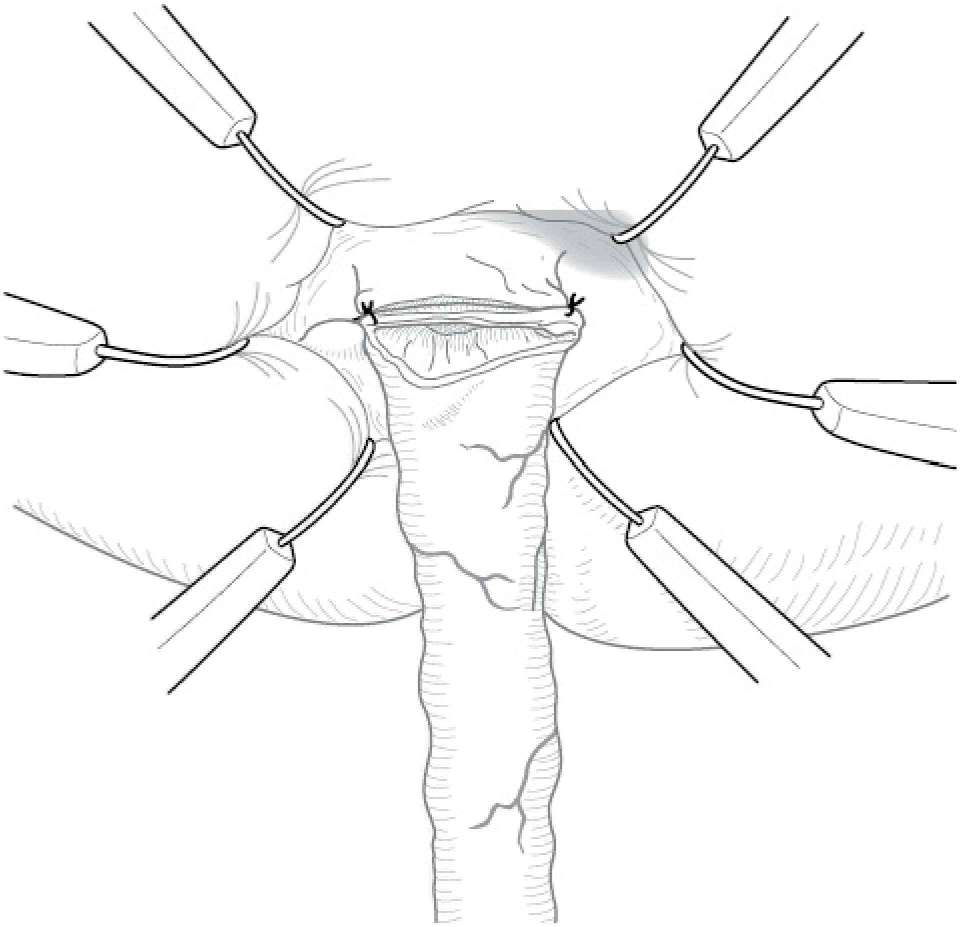
47.30

A key point is to split the muscular tube down to the top of the internal sphincter in the posterior midline (**Figure 47.31**). This splitting prevents the muscular cuff from creating a relative obstruction to the passage of stool. Although dissection of mesenteric vessels is easier with the laparoscopic approach, for rectosigmoid Hirschsprung disease, dissection of many of these vessels can be carried out via the transanal route. Biopsies are sent to determine the level of ganglionic bowel.



47.31

Completion of the anastomosis is very straightforward. The pull-through segment is tacked by the seromusular layer 1–2 cm above the anastomosis in four quadrants with care to not damage anterior structures (**Figure 47.32**). The anastomosis is completed in the standard fashion.



47.32

Swenson procedure

This technique was originally described by Swenson and Bill in 1948 and was the first successful method of treatment for children with Hirschsprung disease. They based their technique on the principle that the diseased portion of the bowel was the aganglionic distal rectum, and that removal of this segment was necessary to allow for normal stooling. The initial incidence of postoperative enterocolitis was fairly high (early 16% and late 27%), and this was attributed to leaving too much aganglionic rectum. The procedure has since been modified by Swenson by creating an oblique anastomosis, in which one resects virtually all of the posterior rectal wall (and some internal sphincter) while leaving 1.5–2 cm of anterior rectal wall.

The technique demands meticulous dissection of the rectum down to within 2 cm of the dentate line. If the dissection moves off the rectal wall, a significant incidence of injury to the genitourinary innervation may occur. Properly performed, the results with this procedure are quite good. However, because of the technical difficulties of the dissection and fear of the associated complications, it had fallen into relative disfavor. With advancement in the understanding of pelvic anatomy and a technique that stresses the importance of proximity to the rectal wall, a transanal Swenson-like pull-through is the operation of choice for some surgeons now. Only experienced surgeons should perform this technique as the risk of nervous injury remains high.

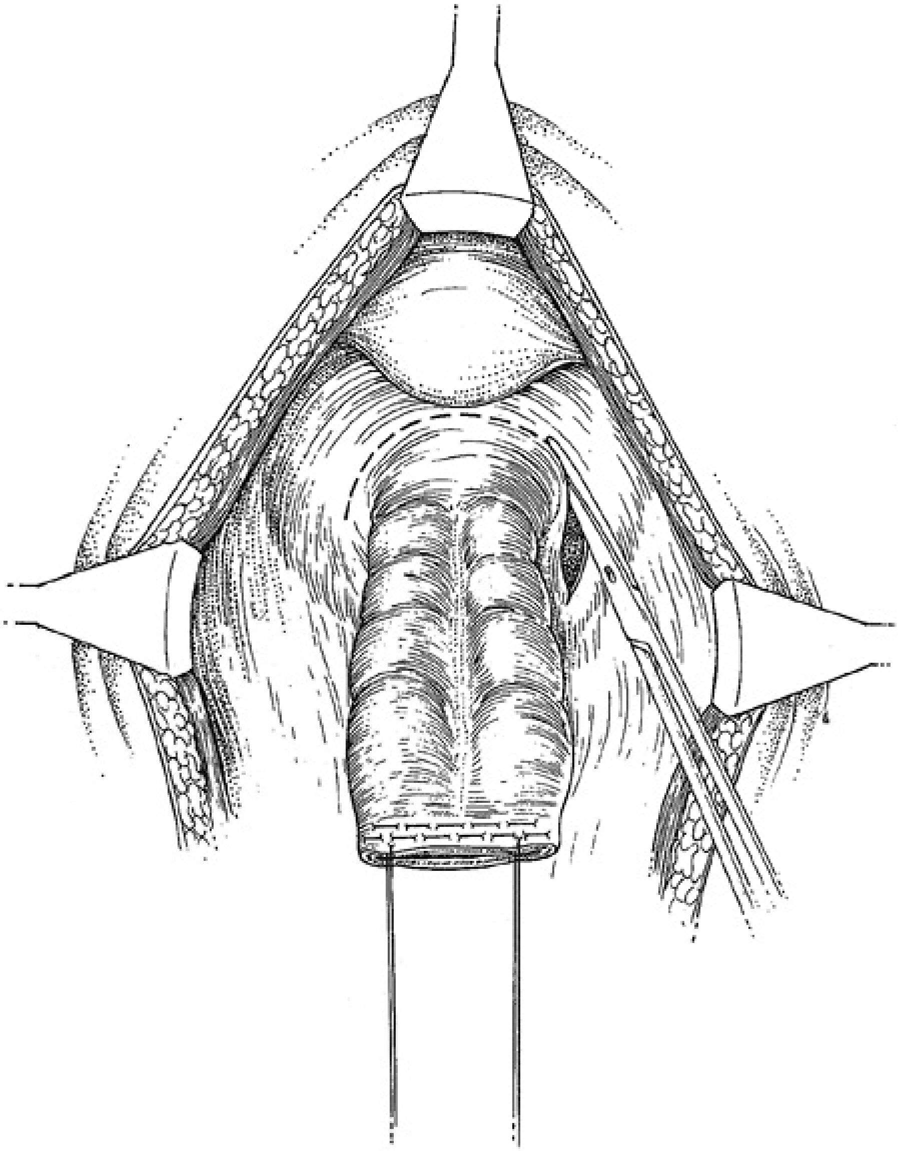
PREOPERATIVE

There is variability in practice and active discussion among pediatric surgeons as to whether or not the child is admitted the day before surgery for a routine bowel preparation. Assessment and preparation are similar to those used in other pull-through operations.

OPERATIVE TECHNIQUE

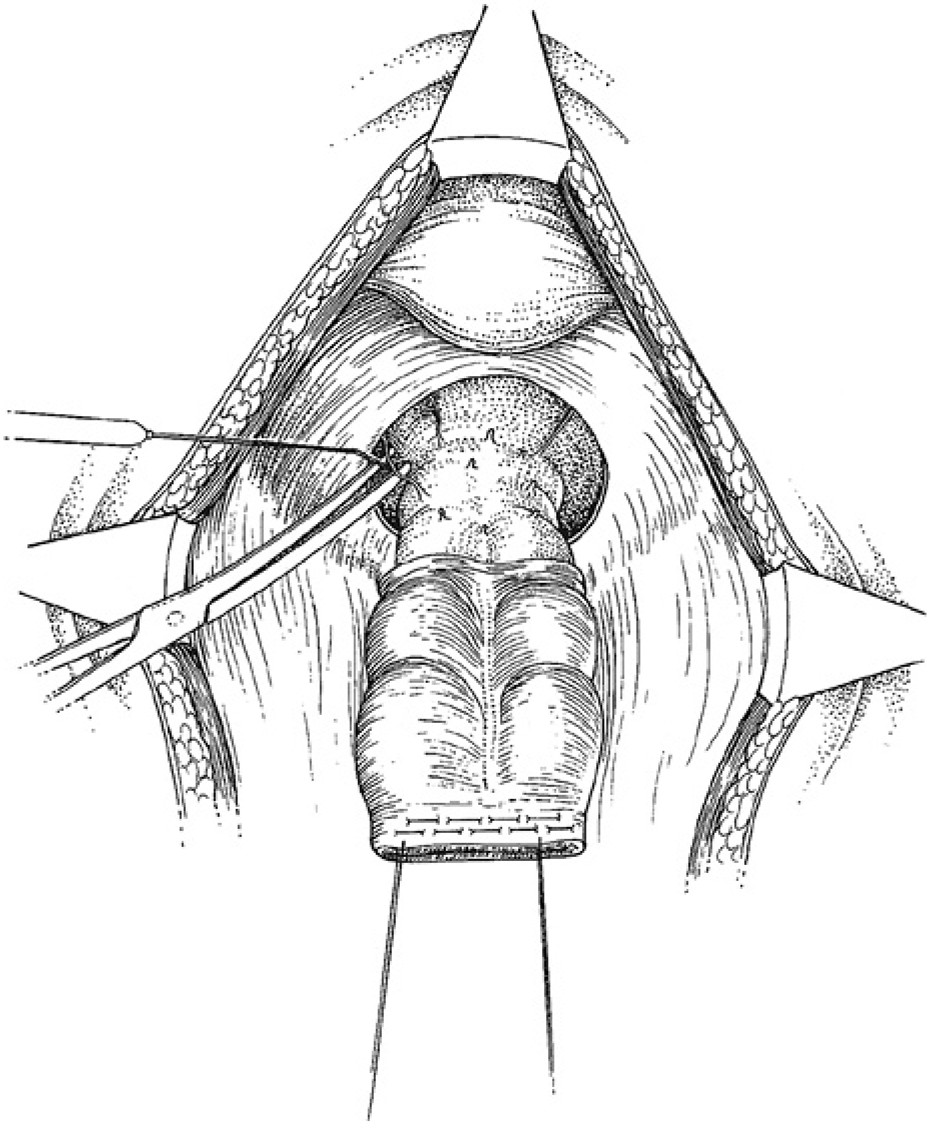
The child is positioned in a fashion similar to that for an endorectal pull-through procedure. The mobilization can begin laparoscopically as previously described. For open procedures, the incision was classically described as a left paramedian incision with take down of the colostomy; however, the modified hockey-stick incision will work equally well.

The redundant aganglionic rectum is excised and proximal ganglionated colon mobilized past the splenic flexure, if necessary, as in the other two techniques. At this point, the peritoneal reflection over the rectum is incised (**Figure 47.33**).



47.33

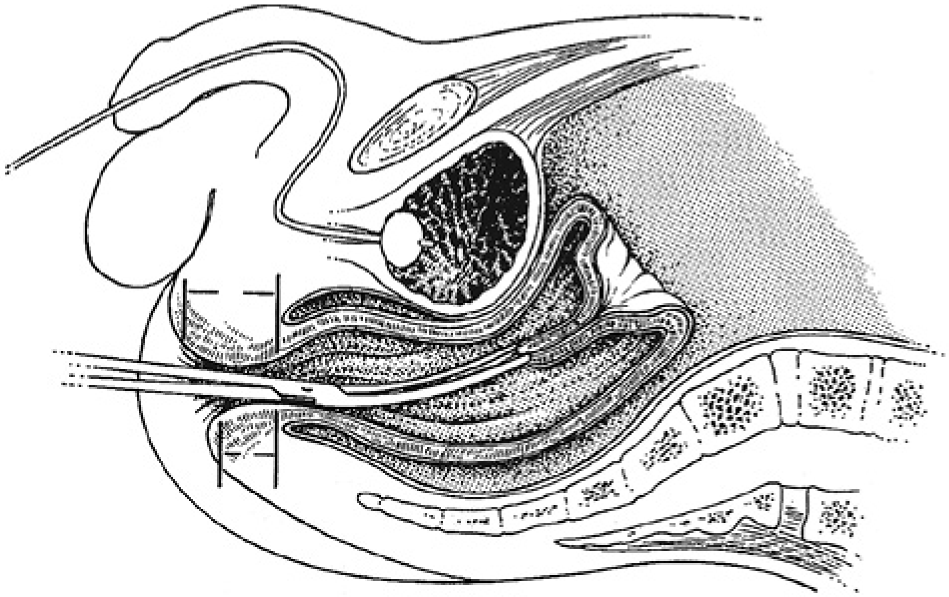
The operating surgeon then dissects the rectum caudally. This is a critical dissection, which demands the surgeon to stay directly on the bowel wall. Dissection is facilitated by the first assistant applying upward traction on the end of the aganglionic rectum. Multiple blood vessels enter directly into the bowel wall; each must be dissected out and can usually be coagulated. Dissection is carried down toward the anal verge, but is not carried as far anteriorly in order to avoid autonomic nerve injury (**Figure 47.34**).



47.34

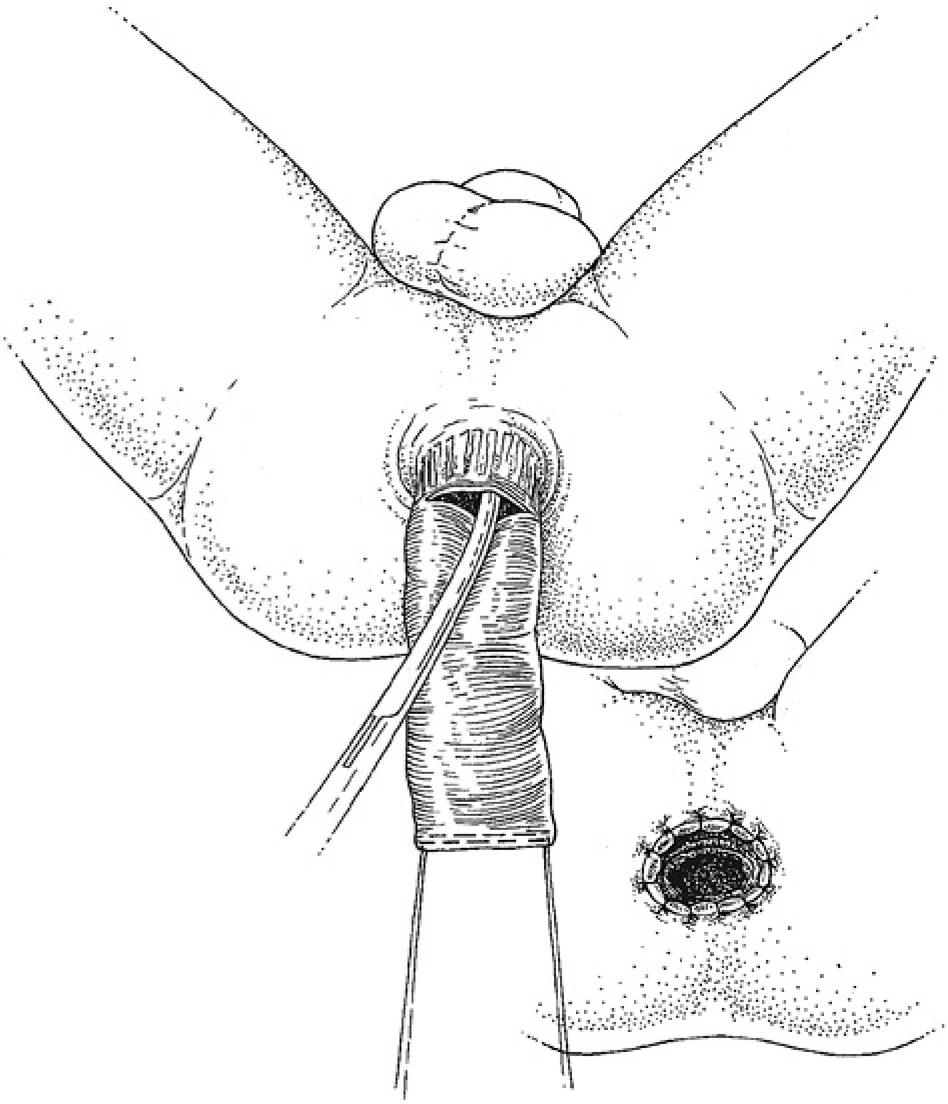
This dissection may be carried out with equal ease via a laparoscopic approach. Deeper pelvic exposure will often require an extra retractor lifting up the bladder, or an externally placed suture which grasps the bladder.

The perineal part of the operation is then started. Should the surgeon feel the dissection is low enough, the rectum is everted with the use of a long clamp (**Figure 47.35**). The anterior half of the everted rectal wall is cut 1.5–2 cm proximal to the anodermal junction. Ultrasound can be used on the everted rectum to ensure internal sphincters are preserved. The posterior wall will be no longer than 1 cm in length. A gently curved incision, which is shorter posteriorly, is thus created along the anterior half of the bowel.



47.35

The ganglionic colon is pulled through and the anastomosis is virtually identical to that for the endorectal pull-through operation (**Figure 47.36**). Classically, silk sutures have been used, but the authors have utilized polyglactin sutures with good results.

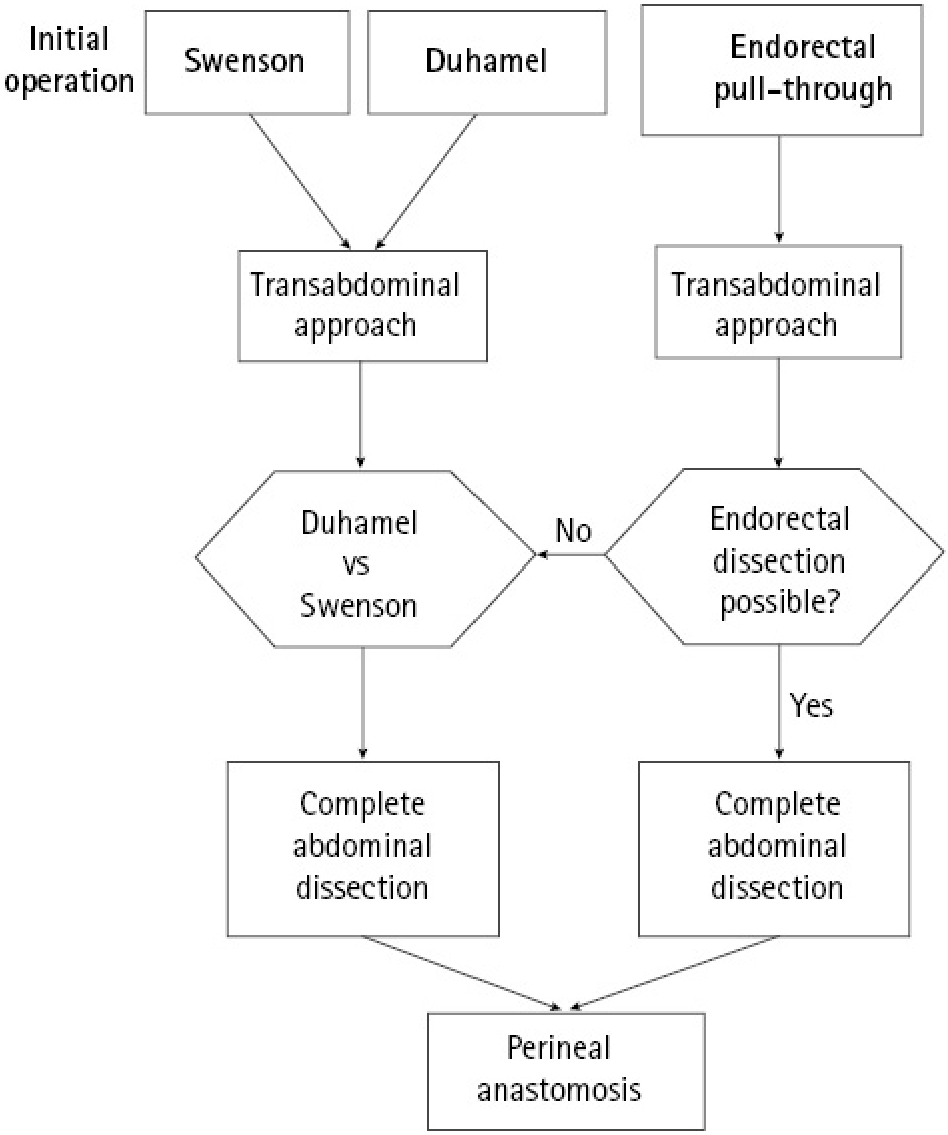


47.36

Alternatively, the set-up for the transanal dissection previously described can be done (see **Figure 47.28**). Starting 0.5–1 cm from the dentate line, a full-thickness mobilization can be performed with the dissection kept on the rectal wall including no perirectal fat. This is essentially a transanal Swenson type procedure. The peritoneal cavity will be entered after a few centimeters of rectum is mobilized. The previously mobilized rectum and sigmoid can then be externalized through the anus. In either method, the anastomosis is allowed to recede and is gently pulled upward from the perineum. Closure is essentially the same as for the previous procedures.

Second pull-through

A child with an initial unsuccessful pull-through operation may present with severe constipation or significant incontinence. These children should undergo a thorough investigation of the details of the initial operation, as well as a review of the pathologic specimens (**Figure 47.37**). This should be done at a referral center with an experienced team. In most cases, an appropriate pull-through has been performed and an aggressive bowel program needs to be instituted. It is essential to rule out a retained segment of aganglionic bowel. Should this be the case, a second pull-through operation will usually be necessary. Because of its relative ease, a Duhamel is a good option. The authors quite commonly perform an endorectal pull-through in patients who have previously undergone this procedure. For those patients who underwent an initial Duhamel procedure, a Swenson pull-through should be used. For children who underwent an initial Swenson procedure, one could also successfully perform an endorectal dissection. If the patient has a stricture in part of the pull-through, one will generally be better off using a Duhamel approach for the "redo" pull-through. Other anatomic problems that should be evaluated include an obstructing Soave cuff, Duhamel pouch or "spur," a twisted pull-through, or a retained or acquired aganglionic segment.


Initial operation
Swenson
Duhamel
Endorectal pull-through
Transabdominal approach
Transabdominal approach
Duhamel vs Swenson
No
Endorectal dissection possible?
Yes
Complete abdominal dissection
Complete abdominal dissection
Perineal anastomosis

47.37

For the child with debilitating incontinence, a detailed history should be taken and an examination to rule out encopresis and to assess the degree of anorectal tone. Anal manometry is particularly helpful in this group, as a lack of normal muscular control is generally felt to be a poor indicator of a successful outcome for a repeat pull-through operation. An occasional child in this latter group may best be served by a diverting colostomy or placement of an appendicostomy tube.

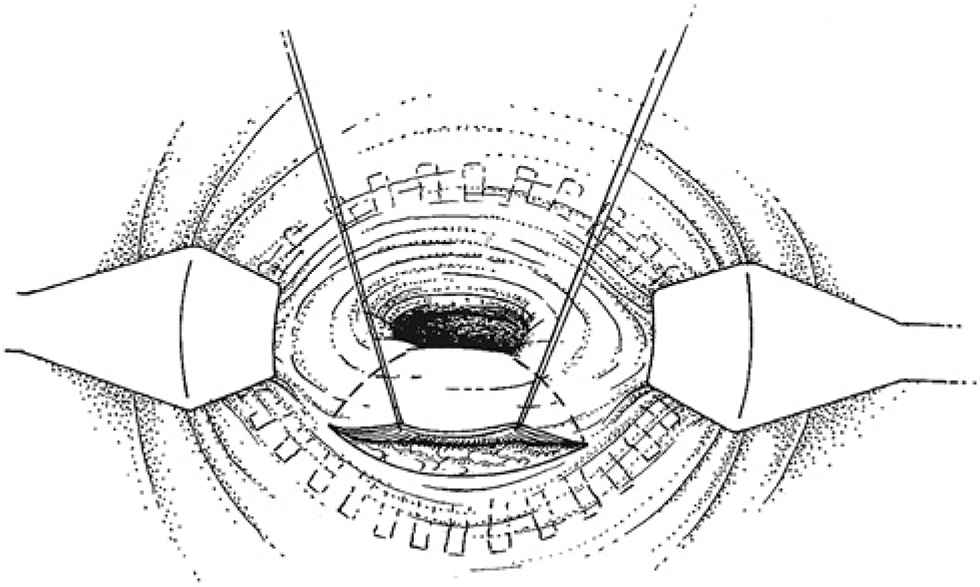
Ano/rectal myectomy

A child with short-segment Hirschsprung disease may be a candidate for an anal/rectal myectomy. The advantage of this procedure is that it avoids an abdominal operation. The procedure must be confined to very short-segment (4 cm or less including the transition zone) Hirschsprung disease. One must be certain that the myectomy is extended beyond the level of the transition zone. Because performing an inadequate myectomy may adversely affect the outcome of a subsequent pull-through operation, a myectomy should be avoided if there is any uncertainty as to the level of disease. More commonly, an ano/rectal myotomy or myectomy is performed for a child who, following a pull-through procedure, has persistent enterocolitis symptoms, or who has difficulty stooling after a pull-through procedure. The most satisfying results with a myectomy are for those children with long-standing enterocolitis after a pull-through procedure.

TRANSANAL APPROACH

The child is placed in an identical position to that for the full-thickness rectal biopsy.

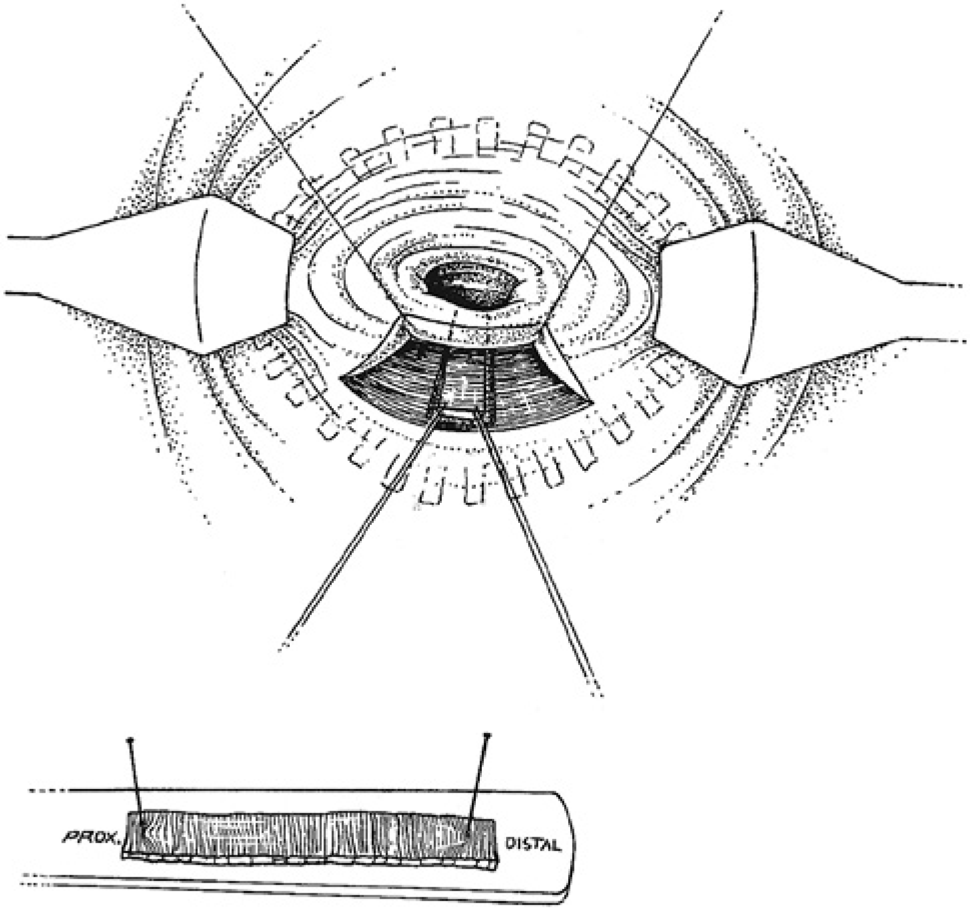
Digital dilatation is performed, and two narrow anal retractors are inserted and held by assistants. The posterior aspect of the dentate line is identified. A 2 cm transverse incision through the mucosa and submucosa starting 0.5–1 cm above the dentate line is made. Following this, a submucosal dissection is carried upward for several centimeters. The mucosal/submucosal flap is held up with silk sutures and can be extended proximally with vertical incisions on either side (**Figure 47.38**).



47.38

The myectomy is then performed by sharply incising the full thickness of the muscle layer and a 0.5–1 cm wide muscle strip is created in the midline. One should try to avoid the use of Bovie cautery until the specimen is obtained—preventing cautery artifact which may make pathologic interpretation difficult. The initial strip removed should be at least 5 cm in length.

Before transecting this strip, two silk sutures are placed proximal to the point of transection so that, should further dissection be necessary, the proximal muscle will not retract beyond the view of the surgeon. The strip should be mounted on a tongue depressor with mounting pins or suture; proximal and distal orientation must be clearly depicted (**Figure 47.39**).

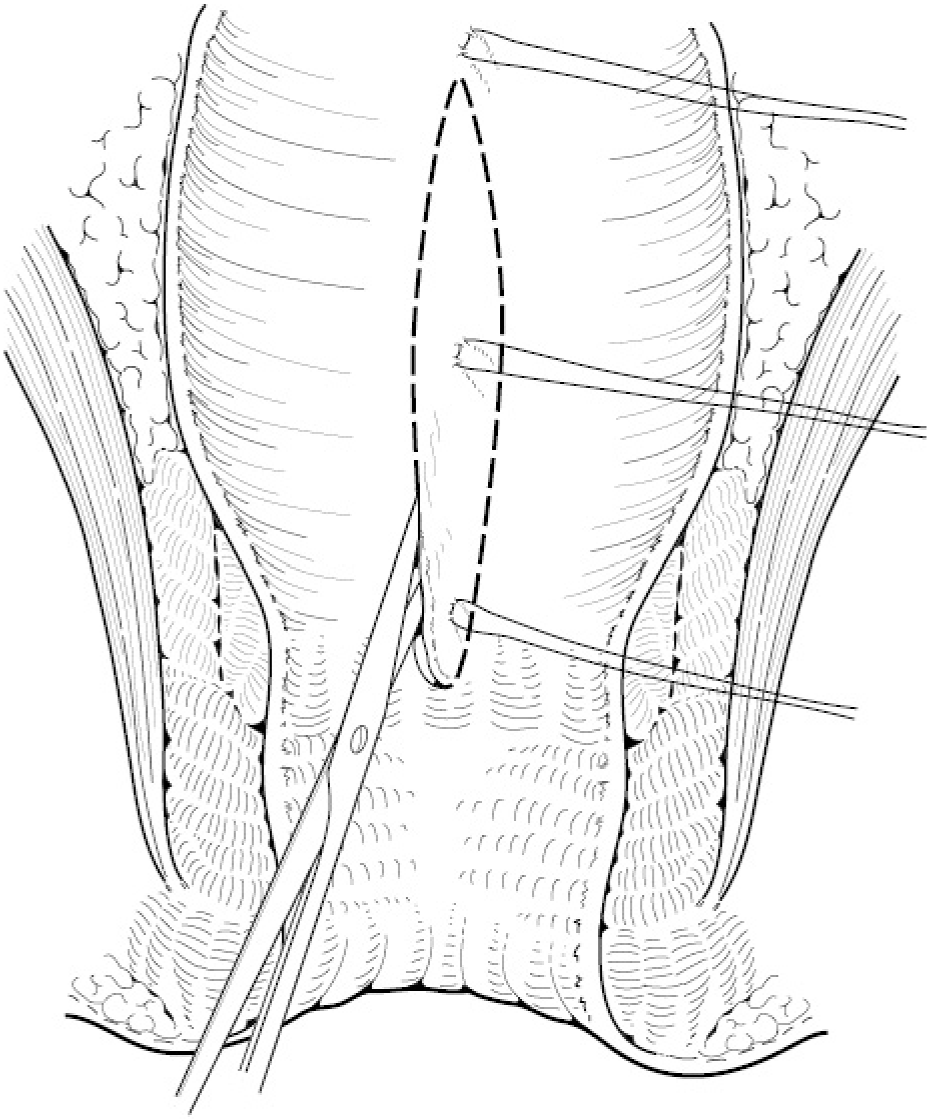


47.39

A frozen section confirming ganglion cells at the proximal margin must be obtained before the procedure can be terminated. The dissection can be carried out in this manner for approximately 4–6 cm. If the surgeon suspects that the level of aganglionosis is longer than 4 cm, it may be advisable to forgo this approach.

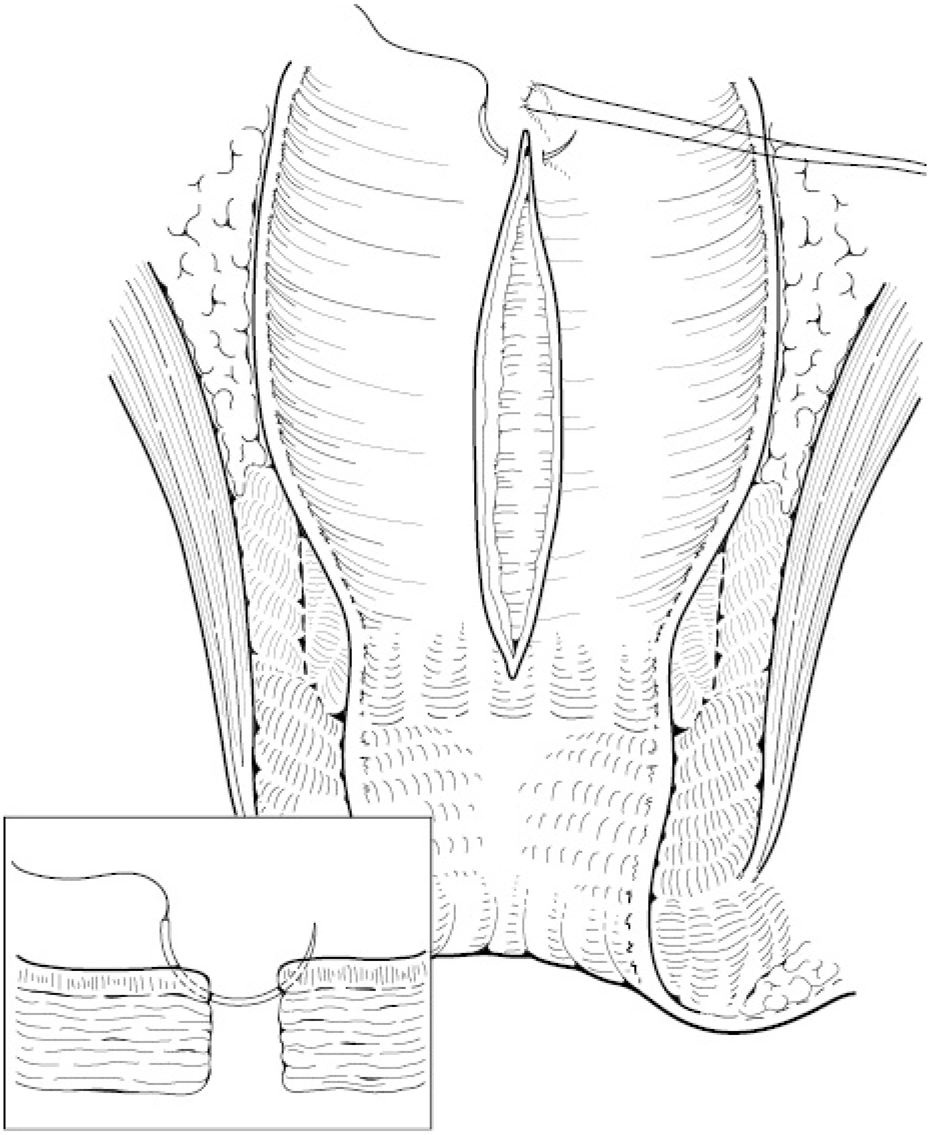
Once a sufficient length of muscle has been removed, hemostasis is achieved with cautery. The wound is irrigated and closed with fine, interrupted, absorbable sutures approximating the mucosa/submucosal dissection.

For patients who have had several procedures, obtaining a submucosal plane may be quite difficult and it is simpler to perform a vertically oriented, elliptical, full-thickness excision (**Figure 47.40**).



47.40

Closure is performed with a running absorbable suture that approximates only the mucosa and submucosa, leaving a defect in the muscular layer (**Figure 47.41**).

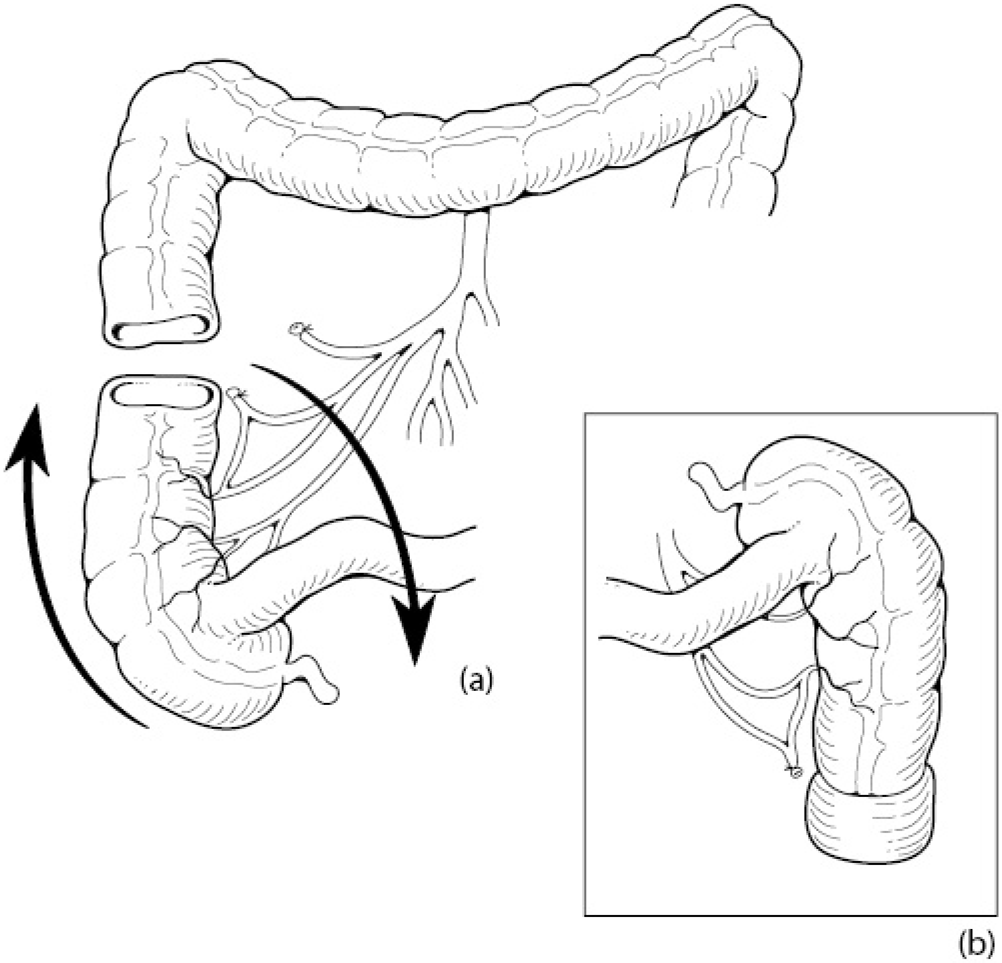


47.41

RIGHT-SIDED AGANGLIONOSIS

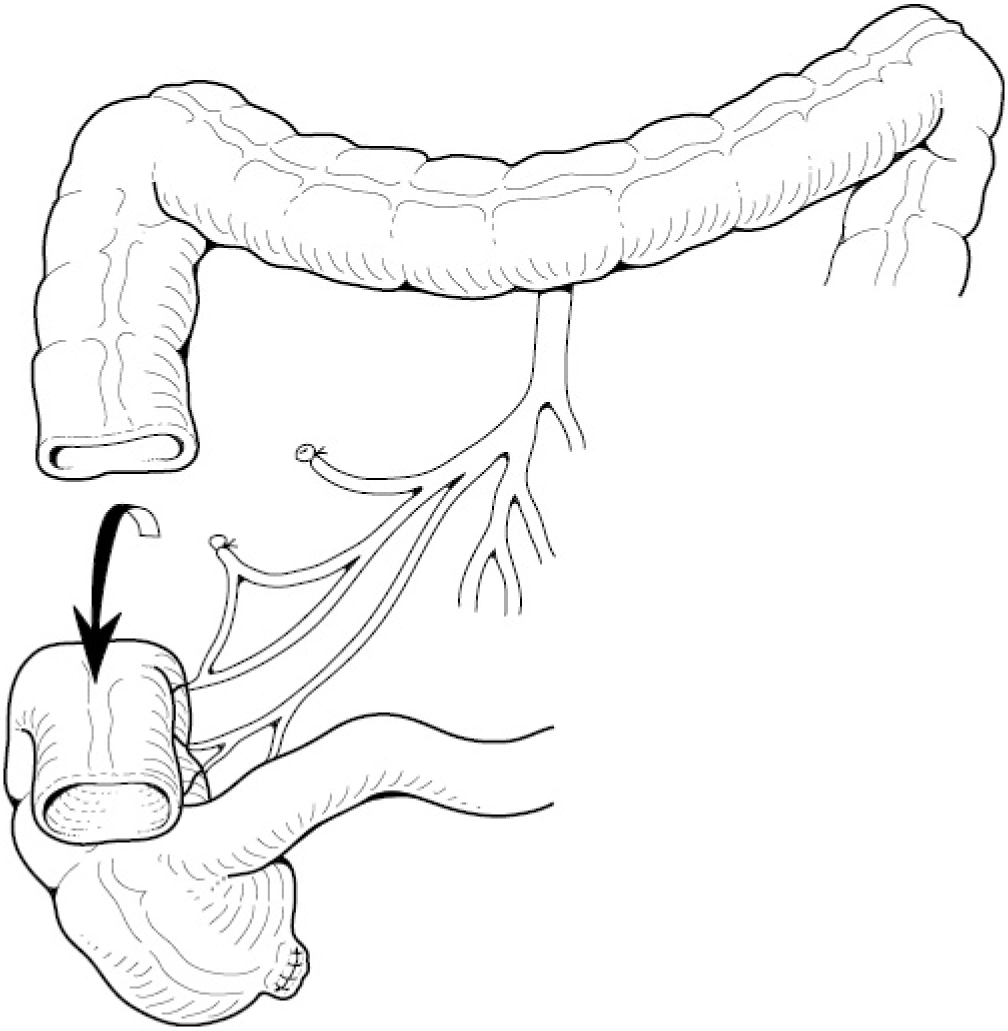
Pull-through procedures in patients with extended colonic disease (to the right of the middle colic vessels) often create problems in maintaining the colon in the correct orientation and are at greater risk for torsion of the pulled-through intestine. This problem was recognized by Duhamel early on.

This problem may be best approached by rotating the right colon in a counterclockwise fashion, as shown in **Figure 47.42a and b**. Because the small bowel then exits the colon from the patient's left side, it is sometimes necessary to follow this with a complete release of the ligament of Treitz (i.e. create a malrotation).



47.42a,b

Alternatively, the colon may be flipped anteriorly (**Figure 47.43**), with reasonably good results.



47.43

POSTOPERATIVE CARE

Following any of the pull-through procedures, if a nasogastric tube was used it can be removed on the first postoperative day. Feedings are begun once gastrointestinal activity returns. The Foley catheter can be removed on the second postoperative day following the endorectal and Duhamel procedures because bladder innervation has not been affected by the pelvic dissection. However, after the Swenson pull-through or redo pull-throughs, the Foley catheter should be left in for 3–5 days because of the possibility that there may be some bladder atony from the dissection. Antibiotics are continued for 24 hours after surgery. Most patients are discharged by the third to fifth postoperative day.

Great emphasis is placed on examining the perineal region for the development of erythema or cellulitis, as this is an early sign of an anastomotic leak. Investigation of a leak consists of an enema contrast study or a computed tomographic scan with contrast medium. Although it is fairly uncommon, if a leak is found, urgent placement of a diverting colostomy is needed.

Parents are given detailed instructions on perineal care and the potential development of enterocolitis. To avoid perineal excoriation, the parents are instructed to apply a thick coat of a zinc oxide-based ointment with each diaper change. If a significant rash develops, more intensified applications are used.

Digital or small Hegar calibrations can be performed safely, starting on the third week after surgery. These are occasionally needed after the endorectal pull-through procedure, particularly in neonates. These dilatations are usually sufficient to prevent anastomotic stricturing. Many surgeons do not routinely dilate, but the anastomosis should be evaluated within weeks of the operation and dilated if a stricture is recognized.

Stool frequency is generally quite high (7–12 bowel movements per day) immediately after the pull-through operation, but this slowly decreases and is generally normal by 6–9 months.

Children should be followed up for several years by the operating surgeon. Occasionally, some develop intermittent bouts of constipation, occasional soiling, and enterocolitis. Most problems with constipation or soiling can be managed with changes in diet or enema regimens. Episodes of enterocolitis are managed with oral metronidazole; however, severe cases will necessitate hospital admission, IV antibiotics, and rectal washouts. As a prophylactic method of avoiding enterocolitis, rectal washouts can be initiated after the first postoperative visit and assessment of the anastomosis, and these may be continued for the first few months following the surgery.

With regard to children with total colonic Hirschsprung disease, large volumes of stool and electrolyte losses can occur after either a modified

Duhamel or an ileoanal endorectal pull-through. These losses must be replaced and are usually controlled with dietary changes and the addition of an opioid agent (e.g. loperamide). Perineal excoriation is very common in these infants and demands constant attention. Normalization of stooling will take much longer than in children with classic Hirschsprung disease, and these children may sometimes require parenteral nutritional support before full enteral feeding can be achieved.

OUTCOME

There is controversy concerning which procedure yields the best results. Long-term outcomes with the Duhamel, Soave, and Swenson techniques are basically similar, provided they are performed with meticulous technique. Depending on the anatomy and history of the patient, the surgeon should be familiar with all of these techniques but should use one as the primary procedure. This is the only way that consistently good results can be achieved.

Of equal importance is the need to follow such patients over long periods of time, as many of the complications (e.g. enterocolitis, constipation, and urgency) may not become manifest until much later in the patient's life.

ACKNOWLEDGMENTS

This chapter is in memory of Daniel H. Teitelbaum MD. Dr Teitelbaum was a professor of surgery at the University of Michigan after completing his training in pediatric surgery at Ohio State University. He dedicated his clinical and research career to the care of children with colorectal disorders and had a tremendous impact on countless children and families affected by Hirschsprung disease.

Illustrations were drawn by Paul Richardson and Shayne Davidson. **Figures 47.19**, **47.21**, and **47.24** are reproduced from Coran and Weintraub, *Surgery, Gynecology and Obstetrics* 1976; 143: 277–82, with permission of the publishers.

VIDEOS

OPS_8e_video_47.1 Laparoscopic biopsy and rectal mobilization
OPS_8e_video_47.2 Suturing laparoscopic biopsy
OPS_8e_video_47.3 Mobilization of sigmoid
OPS_8e_video_47.4 Creation of retro-rectal channel

FURTHER READING

Boley SJ. New modification of the surgical treatment of Hirschsprung's disease. *Surgery* 1964; 56: 1015–17.

Coran AG, Weintraub WH. Modification of the endorectal procedure for Hirschsprung's disease. *Surg Gynecol Obstet* 1976; 143: 277–82.

El-Sawaf MI, Drongowski RA, Chamberlain JN, Coran AG. Are the long-term results of the transanal pull-through equal to those of the transabdominal pull-through? A comparison of the two approaches for Hirschsprung disease. *J Pediatr Surg* 2007; 42: 41–7.

Georgeson KE, Cohen RD, Hebra A, et al. Primary laparoscopic-assisted endorectal colon pull-through for Hirschsprung's disease: A new gold standard. *Ann Surg* 1999; 229: 678–82.

Kim AC, Langer JC, Pastor AC, et al. Endorectal pull-through for Hirschsprung's disease – a multicenter, long-term comparison of results: Transanal vs transabdominal approach. *J Pediatr Surg* 2010; 45: 1213–20.

Langer JC, Durrant AC, de la Torre ML, et al. One-stage transanal Soave pull-through for Hirschsprung disease: A multicenter experience with 141 children. *Ann Surg* 2003; 238: 569–76.

Levitt MA, Hamrick MC, Eradi B, et al. Transanal, full-thickness, Swenson-like approach for Hirschsprung disease. *J Pediatr Surg* 2013; 48(11): 2289–95.

Swenson O, Bill AH. Resection of the rectum and rectosigmoid with preservation of the sphincter for benign spastic lesions producing megacolon. An experimental study. *Surgery* 1948; 24: 212–20.

Wildhaber BE, Pakarinen M, Rintala RJ, et al. Posterior myotomy/myectomy for persistent stooling problems in Hirschsprung's disease. *J Pediatr Surg* 2004; 39: 920–6.

48

Inflammatory bowel disease

AIMEE G. KIM, SAMIR K. GADEPALLI, AND JAMES D. GEIGER

INTRODUCTION

Surgery in Crohn's disease is an adjunct in the repertoire of therapies aimed to restore normal growth and development while preserving bowel length and function as much as possible for as long as possible. Surgery in ulcerative colitis — total proctocolectomy and various restorative techniques — has evolved with ileal pouch–anal anastomosis, resulting in excellent functional outcomes and quality of life.

HISTORY

Both Crohn's disease (CD) and ulcerative colitis (UC) have been described in medical literature for centuries. The first complete description of CD is attributed to Giovanni Battista Morgagni (1682–1771), the "father of anatomic pathology,"[1] in the description of a 20-year old with a long-standing history who died with a perforated terminal ileum. The eponym Crohn's disease (or Crohn disease) is based on an article published in the *Journal of the American Medical Association* in 1932 describing "regional ileitis."[2] Physician Sir Samuel Wilkes coined the term "ulcerative colitis" in a case report from 1859, and similar cases described by Sir William Hale White in 1888 corroborated the condition. The first pediatric cases of UC were described in 1923 in children aged between 8 and 15. Historically, evolution in the medical and surgical management of inflammatory bowel disease (IBD) required much trial and error. Recently however, a growing understanding of the role of molecular biology and immunology in IBD has led to the development and application of biologics (monoclonal antibodies and similar) against specific cytokines and cytokine activity. Nonetheless, patients with IBD often necessitate surgical management, and a variety of procedures have been developed and honed with the goals of maximal preservation of bowel length, function, and quality of life. These include stricturoplasty, bowel resection and anastomosis, various temporizing and permanent ileostomies, and partial and complete colectomy with several approaches for bowel continuity. The pediatric patient with IBD presents with challenges to growth, nutritional, and psychologic development that require thoughtful consideration when determining the timing and type of procedure.

PRINCIPLES AND JUSTIFICATION

IBD, characterized by chronic and recurrent inflammation of the gastrointestinal tract, consists of two main subtypes: Crohn's disease and ulcerative colitis. The incidence of pediatric IBD is approximately 10 per 100 000 children in the United States, and rising.[3] The pathological and clinical manifestations of the diseases are generally similar to those described in the adolescent/adult population. Multifactorial genetic, environmental, and microbial influences to the pathophysiology are still being characterized.

CD can involve any portion of the gastrointestinal tract in a discontinuous transmural inflammatory process, and can be further subdivided into inflammatory (non-stricturing, non-penetrating), stricturing, penetrating, and combination subtypes. A quarter of patients with pediatric-onset CD will require operative resection within 5 years of diagnosis, and, of these, ileocecectomy is the most frequently performed surgical intervention (53%).[4] A description of a laparoscopic-assisted ileocecectomy is included in the "Operations" section. Surgical management of clinically significant strictures has evolved to minimize bowel resection (and reduce the risk of developing short bowel syndrome) and maximize bowel function utilizing a modification of the Heineke-Mikulicz procedure for pyloroplasty. A key point is ensuring that all of the small and large bowel has been carefully inspected. Contraindications to stricturoplasty include, for example, the occurrence of several small strictures in close proximity (better managed with a resection) and association of a stricture with a fistula or abscess (bowel segment should be excised). Perianal disease is also a common cause for morbidity in CD, affecting 20% of patients[5] and often requiring surgical intervention for management of recurrent painful abscesses and fistulas. Severe perianal disease refractory to both local surgical and escalating medical management may be improved with creation of temporizing diverting ileostomy.

UC is characterized by continuous inflammation of the colon, extending proximally from the rectum, and is limited to the mucosa. UC in children will present with extensive pancolitis in 60–80% of cases, with a more severe disease course compared to that of adults, and a 30–40% colectomy rate at 10 years.[6] Not surprisingly, the rate of adenocarcinoma is also higher in childhood-onset UC than that in adult-onset UC: Cumulative probabilities of developing colorectal carcinoma are 5.5, 10.8, and 15.7% at 10, 20, and 30 years after onset of UC.[7] Total proctocolectomy can be curative in the patient with UC,[8] but, as described below, it can be challenging to ascertain that a child with colonic disease truly has UC. Historically, patients with UC would undergo total proctocolectomy with permanent end ileostomy. Since the 1940s, several approaches to restoring functional intestinal continuity have been described: Ileorectal anastomosis (IRA; limited to those with only mild-to-moderate disease of the rectum and requiring routine surveillance of remaining rectal mucosa), straight ileoanal anastomosis (SIAA; potentially problematic because of limited reservoir capacity of ileum), and ileal pouch–anal anastomosis (IPAA) also known as restorative proctocolectomy (RP) (the currently recommended standard curative operation for UC). Further, various techniques exist for creation of the ileal reservoir (J-pouch, lateral side-to-side, S-pouch), and the procedure may be performed in one (RP with or without ileal pouch), two (RP with or without ileal pouch and diverting ileostomy, followed by ileostomy reversal), or three stages (Stage 1: Subtotal colectomy with end ileostomy; followed by Stage 2: Completion proctectomy with ileoanal anastomosis with or without ileal pouch and diverting ileostomy; followed by Stage 3: Ileostomy reversal). Most pediatric surgeons favor a three-stage ileoanal anastomosis with creation of a pouch and a temporary diverting ileostomy after coloproctectomy. The most recent modification is the endorectal double-stapled (ERDS) ileoanal pouch anastomosis. This combines the speed and simplicity of double stapling with the reassurance of endorectal dissection allowing reach and clear identification of the low rectum and dentate line while avoiding injury to the parasympathetic and sympathetic nerve of the pelvic plexus, and preventing stretch of the anal sphincters.[9]

The decision to perform a protective ileostomy is highly debatable.[10] The advantage of eliminating the ileostomy is the ability to forgo a subsequent surgery and the potential complications associated with an ostomy. A diverting ileostomy may be omitted in select cases with comparable postoperative outcomes (e.g. stapled anastomosis, minimal tension on the ileoanal anastomosis, intact tissue rings, good hemostasis, and absence of airleak, malnutrition, toxicity, anemia, or prolonged steroid use). More patients with a diverting ileostomy and defunctionalized pouch may develop anastomotic stenosis necessitating dilations.

A few additional notable points about IBD in the pediatric patient are important to the consideration of surgical management. Growth failure is a predominant feature in the presentation of IBD in children, and more severe in CD than in UC. Pediatric CD can often involve only the colon, and small bowel disease may not be apparent until later; further, it is uncommon to have disease limited to the small bowel. Designation of the subtype IBD-unspecified (IBD-U), or indeterminate colitis, reflects the challenge of discriminating between CD and UC in children. Studies examining the subtypes of IBD have found approximately 15% of patients undergo a change in diagnosis[3] and, generally UC and IBD-U diagnoses may later be changed to CD. In cases of severe colonic IBD, IBD-U, or perianal disease related to CD, creation of a diverting ileostomy has been found to be a good temporizing measure for symptom alleviation, clinical stabilization, and growth normalization, and it allows time to clarify a diagnosis.

Characterizing disease progression for the purposes of clinical management, research, and comparison of outcomes after therapy/intervention has been aided by the development and validation of clinical scoring tools: Pediatric Crohn's Disease Activity Index (PCDAI)[11] (**Table 48.1**) and Pediatric Ulcerative Colitis Activity Index (PUCAI)[12] (**Table 48.2**). The PCDAI consists of subjective (symptoms) and objective components (bloodwork, physical examination) to comprise a score from 0 to 100 with higher scores indicating more active disease. A score <10 indicates inactive/mildly active disease and ≥30 reflects moderate disease activity; a decrease of 12.5 points or greater represents significant clinical improvement. PCDAI does not correlate with mucosal inflammation, an important end point in the treatment of IBD. The PUCAI, in contrast, purely symptom-based, with a total score from 0 to 85, has shown good correlation with endoscopic disease activity. Remission is defined as <10, moderate disease is ≥30, and severe disease is ≥65. The minimal important difference (MID), or the smallest change that should provoke a change in management, is 20.

Table 48.1 **Pediatric Crohn's Disease Activity Index (PCDAI)**

HISTORY (Recall, 1 week)	
Abdominal pain:	
None	0
Mild	5
Moderate/Severe	10
Stools: (per day)	
0–1 liquid, no blood	0
≤2 semi-formed w/ small blood or 2–5 liquid	5
Gross bleeding, ≥6 liquid, or noctural diarrhea	10
Patient functioning:	
No limitation of activities, well	0
Occasional difficulty in maintaining age-appropriate activities	5
Frequent limitation of activity, very poor	10
LABORATORY	
HCT (%):	
<10 years	
>33	0
28–32	2.5
<28	5
11–19 years, female	
≥34	0
29–33	2.5
<29	5
11–14 years, male	
≥34	0
30–34	2.5
<30	5
15–19 years, male	
≥37	0
32–36	2.5
<32	5
ESR (mm/hour):	
<20	0
20–50	2.5

Table 48.1 Pediatric Crohn's Disease Activity Index (PCDAI)

>50	5
Albumin (g/dL):	
≥3.5	0
3.1–3.4	5
≤3.0	10
EXAMINATION	
Weight:	
Weight gain or voluntary weight stable/loss	0
Involuntary weight stable, weight loss 1–9%	5
Weight loss ≥10%	10
Height (score “At diagnosis” OR “Follow-up:”	
At diagnosis:	
<1 channel decrease	0
≥1, <2 channel decrease	5
>2 channel decrease	10
Follow-up:	
Height velocity ≥−1SD	0
Height velocity <−1SD, >−2SD	5
≤−2SD	10
Abdomen:	
No tenderness, no mass	0
Tenderness, or mass without tenderness	5
Tenderness, involuntary guarding, definite mass	10
Perirectal disease:	
None, asymptomatic tags	0
1–2 indolent fistula(s), scant drainage, no tenderness	5
Active fistula, drainage, tenderness, or abscess	10
Extra-intestinal manifestations:	
(Fever ≥38.5 for 3 days over past week, definite arthritis, uveitis, *E. nodosum*, *P. gangrenosum)*	
None	0
One	5
≥Two	10
TOTAL (0–100)	

Table 48.2 **Pediatric Ulcerative Colitis Activity Index (PUCAI)**

Abdominal pain:	
No pain	0
Pain can be ignored	5
Pain cannot be ignored	10
Rectal bleeding:	
None	0
Small amount only, <50% of stools	10
Small amount with most stools	20
Large amount (>50% of stool content)	30
Stool consistency of most stools:	
Formed	0
Partially formed	5
Completely unformed	10
Number of stools per 24 hours:	
0–2	0
3–5	5
6–8	10
>8	15
Nocturnal stools (any episode causing wakening):	
No	0
Yes	10
Activity level:	
No limitation of activity	0
Occasional limitation of activity	5
Severe restricted activity	10
TOTAL (0–85)	

PREOPERATIVE ASSESSMENT

When considering operative management for a child with IBD, due diligence on the part of the surgeon is critical. The diagnosis should be as firm as possible — recurrent disease in an ileal pouch with fistula formation is not uncommon in Crohn's colitis that was believed to be UC. Repeat endoscopy with biopsies should be performed to rule out CD in cases of

UC or IBD-U, and the disease process should be fully staged. If biopsies have been performed elsewhere, they should be reread by pathologists at the center where the operation is to be carried out. Imaging must also be reviewed: Specifically, in cases of CD, a contrast upper gastrointestinal series with small bowel follow-through can be helpful, but more recently, cross-sectional imaging with CT or MR enterography is the diagnostic modality of choice (the latter is favored for eliminating radiation exposure and providing more detailed visualization of disease extent and penetrance). Though not always necessary, a contrast enema is commonly performed prior to reversal of an ostomy after RP to evaluate for leak or stricture that would compromise function of the pouch or ileoanal anastomosis. Bloodwork and biomarkers important to ascertaining the nutritional status of the patient and potentially additional risks for surgical complications include CRP, ESR, albumin, fecal calprotectin, as well as drug levels for immunomodulators and biologics. Timing of an operative intervention should be carefully considered with respect to the patient's clinical course, nutritional status (e.g. Does the patient need temporary parenteral nutrition prior to surgery?), medical therapy (e.g. When was the last infliximab dose? Are stress-dose steroids indicated?), and in close discussion with the patient's gastroenterologist and family.

Indications for and implementation of a preoperative bowel prep have been the subject of debate in both adult and pediatric literature. There is no clear Class I evidence to support bowel prep if appropriate broad-spectrum preoperative antibiotics are administered and intraoperative spillage is minimized, but most evidence is generally derived from adult literature.[13] Our group allows for surgeon preference and surgeons may elect to forgo the bowel prep, or choose to prescribe one or both components of a mechanical and antibiotic regimen. The protocol allows for some pediatric patients to be admitted preoperatively specifically for the bowel prep (e.g. under 3 years of age, at risk of dehydration, poor compliance). In general, patients are limited to a clear liquid diet 2 days prior to surgery. The mechanical prep consists of a combination of Miralax® and Dulcolax® administered the day before surgery. The antibiotic prep, also prescribed for the day prior to surgery, consists of three doses each of neomycin (15 mg/kg/dose) and erythromycin (20 mg/kg/dose). Bowel preps must be administered with caution, if at all, in those with unstable IBD and/or obstructive symptoms.

The Enhanced Recovery After Surgery (ERAS) Society guidelines optimize care of the postoperative adult patient to improve outcomes while reducing waste. The recommendations, being slowly trialed and implemented in the pediatric population, also include preoperative elements such as counseling prior to surgery, minimal preoperative fasting, and limited bowel preps. Patients in whom an ostomy is possible should have the skin site marked by an enterostomal therapist before surgery. Data surrounding perioperative stress-dose steroids are lacking in the pediatric population; they should be considered in children who have been on steroid therapy within the past year, especially sustained use at high doses. Though children are generally less at risk for thromboembolic complications compared to adults, IBD is a known risk factor due to the heightened inflammatory state, and, further, many pediatric IBD patients have secondary risk factors such as need for major surgery, presence of a central line, and diminished mobility postoperatively.[14,15] Conservative non-pharmacologic thromboprophylaxis (e.g. hydration, mobilization, pneumatic/compression) should be the norm in all hospitalized children with IBD, and strong consideration should be given for pharmacologic prophylaxis with perioperative low molecular weight heparin. A Foley catheter should be placed if surgery will involve pelvic dissection, as the risk of urinary retention may be higher, but duration should be limited as it may impede early mobility.

OPERATIONS

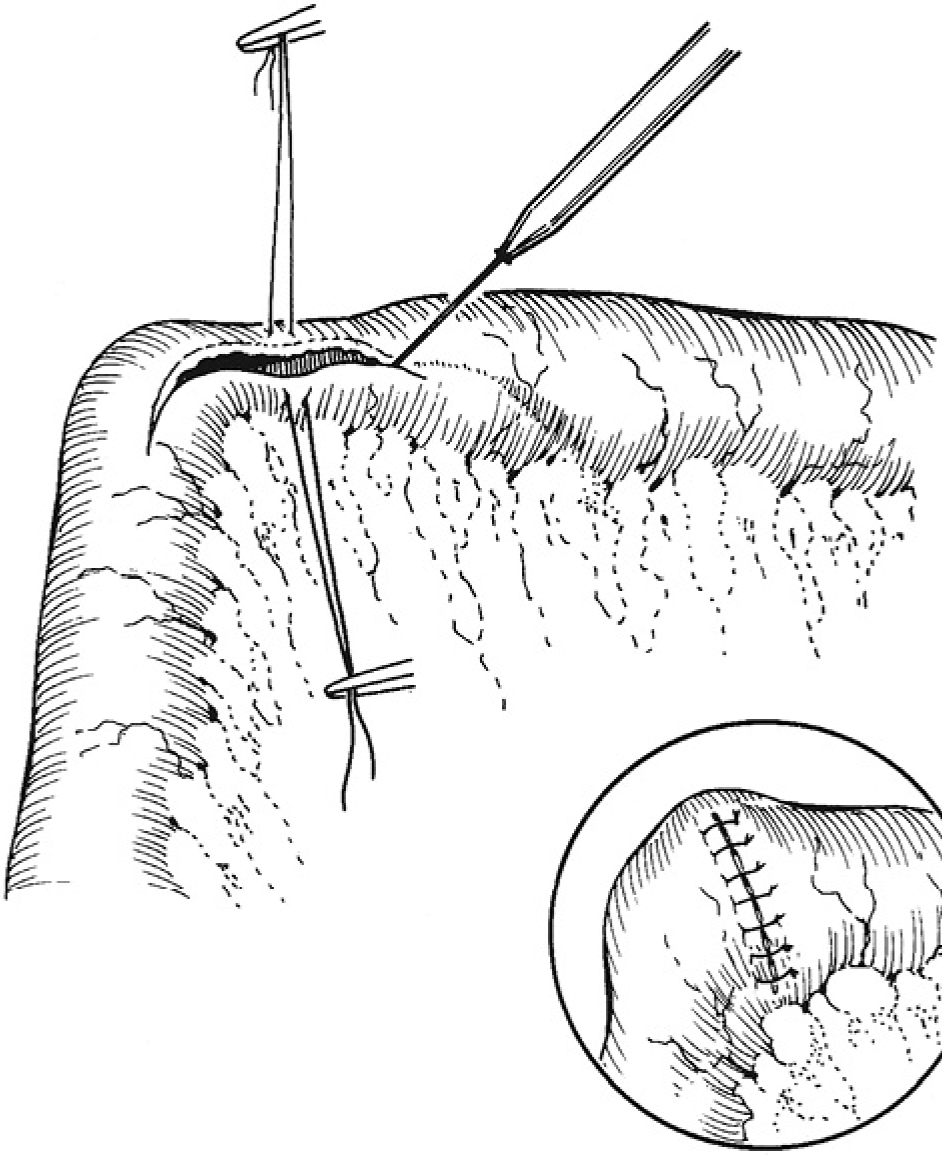
Seton placement

For an abscess that is 1–4 cm in diameter, an incision is placed just over the abscess and at least one counter-incision by the abscess edge and the site is probed and irrigated to break up loculations. A vessel loop is inserted and secured to itself with silk ties. For larger and deeper abscesses, more than two counter-incisions may be indicated and Penrose drains are utilized in lieu of vessel loops. The setons may be left in place for weeks to months even after the abscess is drained, to allow the inflammation to subside and obtain adequate control of their disease. Alternatively, an ultrasound can be used to identify the tract of the fistula using a linear probe placed into the anus. An entry needle can be guided through the fistula pathway using real-time guidance, ensuring accurate placement of the seton(s) for optimal drainage without adjacent injury.

Stricturoplasty

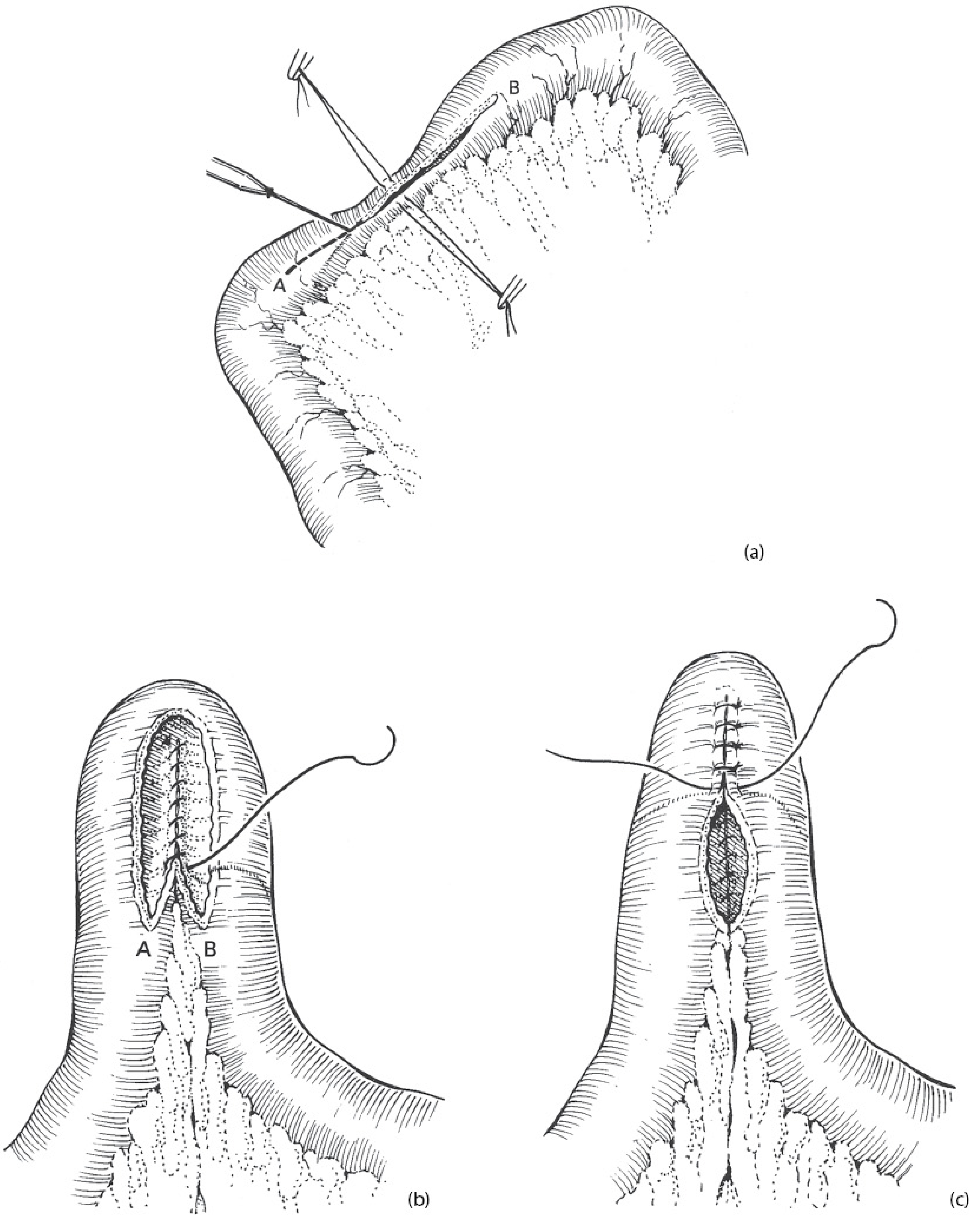
Stricturoplasty may be performed open or laparoscopically. The patient should be positioned supine. Incision and port placement are dependent on location of the strictures, but an initial umbilical port or periumbilical incision allows for greatest versatility, with additional port placements or vertical extension of the periumbilical incision as needed. Upon identification, each stricture is initially marked with a silk suture.

A typical short-segment stricture is managed by placing traction sutures above and below the stricture on the anti-mesenteric surface of the bowel. Using cautery, the bowel is opened longitudinally along the antimesenteric surface. It is then approximated transversely using interrupted 4/0 absorbable or non-absorbable sutures, and a second layer of Lembert sutures is placed (**Figure 48.1**).



48.1

For strictures that are longer, the bowel is folded upon itself so that the stricture is at the apex and opened enough to accommodate a linear stapler with one leg in each of the proximal and distal limbs. When performing the stricturoplasty by and in an open fashion, the stricture at the apex is incised and the incision extended along a portion of both proximal and distal limbs (**Figure 48.2a**).

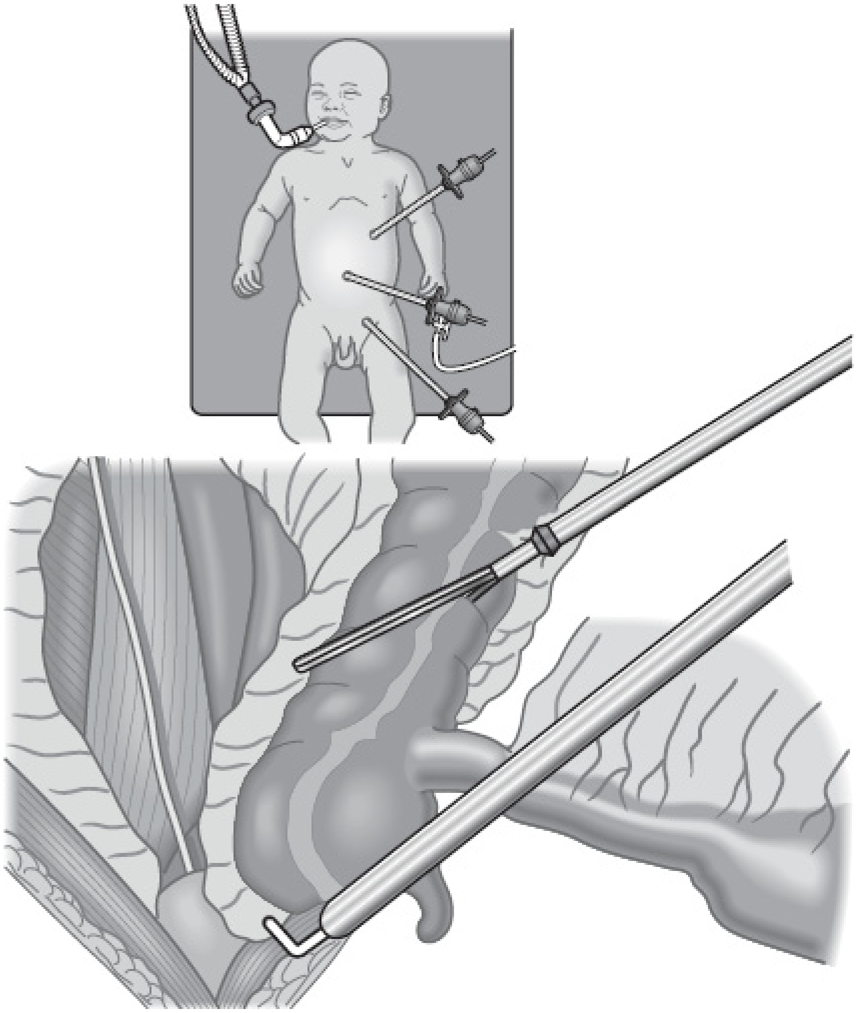


48.2a–c

The linear stapler then staples and divides the tissue between the proximal and distal limbs, creating a common channel with a larger lumen. Alternatively, as shown in **Figure 48.2b and c**, the limbs are anastomosed using an inner layer of running 4/0 absorbable sutures and interrupted Lembert sutures on the outside.

Laparoscopic ileocolectomy

The child should be placed supine. Three or four 5 mm laparoscopic ports are placed in the right upper and left lower quadrants, as well as an umbilical port (**Figure 48.3**). These should allow adequate triangulation of the working ports to facilitate dissection. A fourth port may be needed for additional traction. Prior to beginning the dissection, one should intraoperatively stage the extent of the disease. Running of the small bowel with two atraumatic graspers should be performed looking for other areas of stricture or inflammation.



48.3

The procedure is similar to the open approach, with one working port grasping the bowel with an atraumatic instrument and a hook cautery instrument or scissors used to initially mobilize the right colon and ileum.

Once full mobility has been achieved, two approaches may be taken. First, the bowel can be removed via one of the laparoscopic ports or a right mid-abdomen transverse incision. The incision should be just big enough to allow the inflamed bowel and colon to be eviscerated. The mesentery can be ligated, bowel resected, and anastomosis performed, all extraperitoneally, and the bowel returned to the peritoneum. An alternative approach is the complete laparoscopic excision and intraperitoneal anastomosis. While the latter approach is advocated by surgeons, because the abdominal incision used to remove the specimens is often the same size as the incision for the laparoscopic-assisted approach, the authors do not advocate this technique.

After closure of the peritoneum and fascia, the ports are removed, and incisions are injected with 0.25% Bupivicaine. In general, the 5 mm incisions do not require fascial closure.

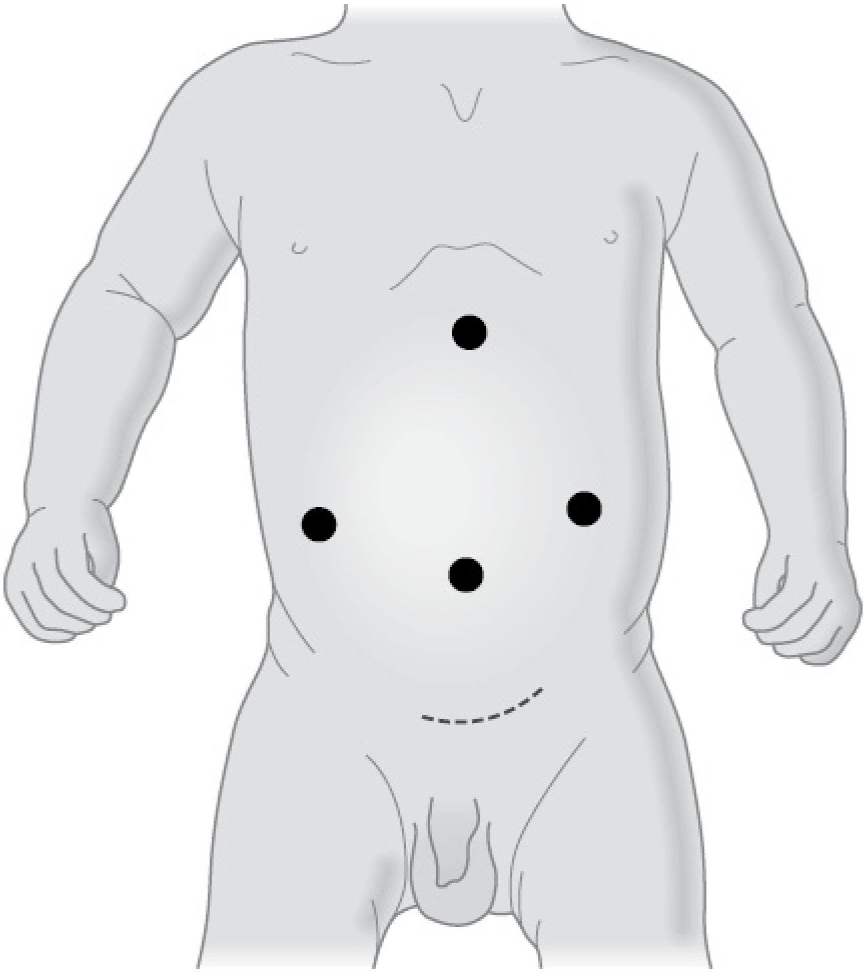
Laparoscopic-assisted total proctocolectomy with end ileostomy

PATIENT POSITIONING AND INCISION

The child is placed supine in staged surgery. In single-stage or restorative stage surgery, the majority of the procedure can be done with the child in the supine position, with the legs placed in previously aligned stirrups for the pull-through and transanal anastomosis portion. In lithotomy, care should be taken to avoid pressure on the posterior portion of the leg with leg supports for older children and skis for smaller children. Careful padding of the lower extremities is critical to avoid neurovascular injury.

TECHNIQUE

The approach to the laparoscopic-assisted proctocolectomy consists of a Pfannenstiel incision just above the pubic symphysis (made either now or at the time of removal of the colon from the abdominal cavity) and four 5 mm ports as shown in **Figure 48.4**: At the umbilicus initially for insufflation and then camera with a 30-degree telescope, at the epigastrium, at the proposed ostomy site (right lateral abdomen), and in the left lateral abdomen.



48.4

For the Pfannenstiel incision, the skin incision is made with a #15 blade scalpel and subcutaneous tissue is dissected with electrocautery to the level of the fascia. Fascia is incised transversely and subfascial flaps are raised cephalad to the level of the umbilicus. The peritoneum is entered via a vertical incision.

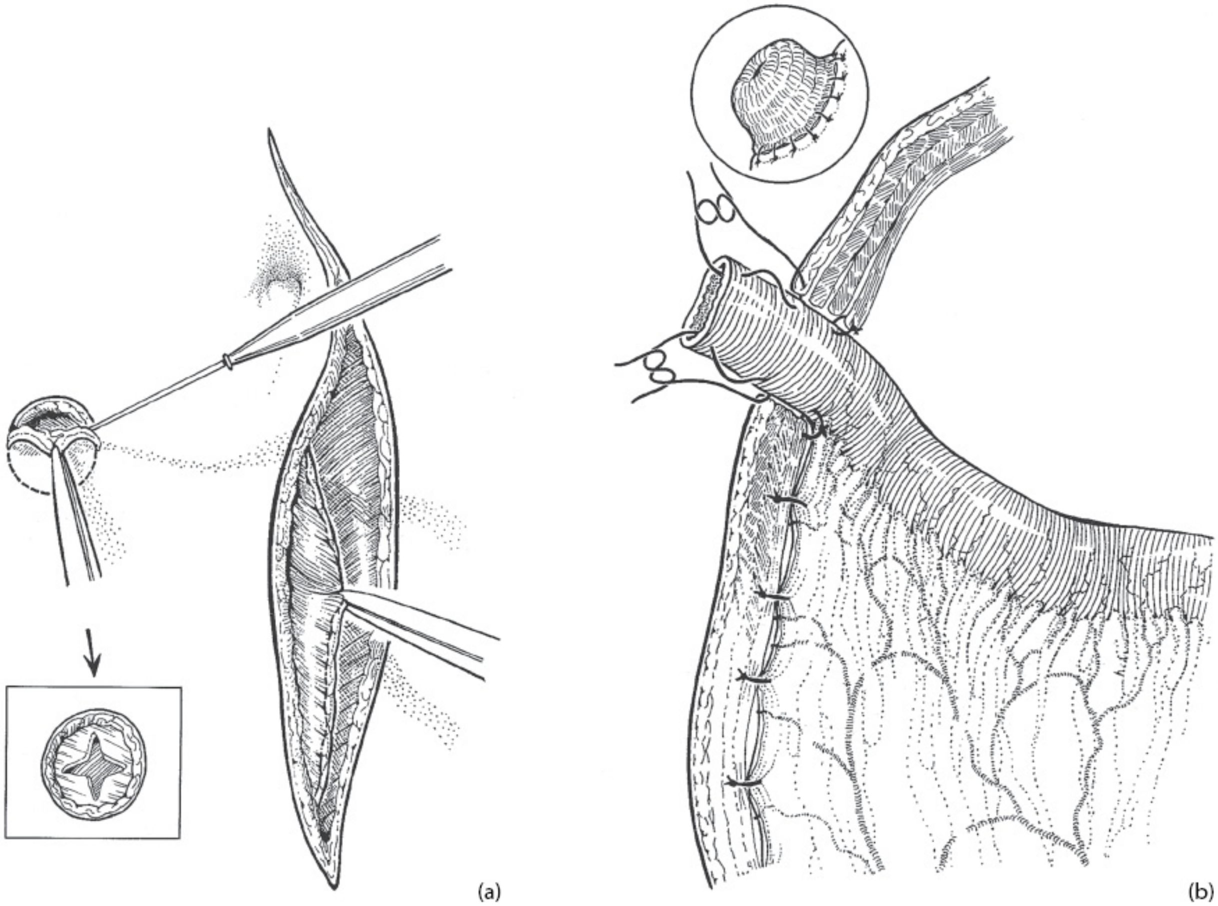
The colon, from the cecum toward the sigmoid colon, is mobilized and dissected utilizing a medial to lateral approach, taking down the mesentery first, followed by the lateral peritoneal attachments. Care is taken to avoid injury to the duodenum at the hepatic flexure or placing traction on the spleen at the splenic flexure. Initial mobilization of the lateral attachments is facilitated by "airplaning" the patient to the contralateral side, with traction on the colon, with a blunt bowel grasper through either the epigastric or the contralateral trocar sites, and using cautery scissors via the remaining trocar site. The ureters are identified early during this dissection. Depending on the site of mobilization, the camera and operating ports will vary.

For mobilization of the omentum off the transverse colon, a pair of 5 mm ultrasonic scissors is helpful, after which the omentum is reflected cephalad. Care is taken to identify and staple or ligate the middle colic vessel. Once the colon and mesentery are fully mobilized, the low transverse suprapubic (Pfannenstiel) incision is made. Alternatively, the Pfannenstiel incision can be made at the beginning and a gel-port placed to be used for a fifth port during the colon mobilization. A wound protector is placed in the Pfannenstiel incision to aid with gentle retraction and visualization. The operating surgeon pulls the terminal ileum (TI) out through the incision and transects the TI approximately 1 cm from the ileocecal valve with an Endo GIA□ stapler (Medtronic, Minneapolis, MN, USA). The entire colon is then eviscerated from the abdomen to the level of the sigmoid colon, and the dissection is carried distally along the sigmoid and rectum, taking care to stay just on the wall of the colon. Just above the peritoneal reflection, the rectum is transected with a blue load of the Endo GIA□ stapler. The colon specimen is sent to pathology. Prolene sutures may be placed at the rectal stump for ease of identification in subsequent surgery. At this point, the ileum is brought up to an end ileostomy.

An alternative approach in a pre-adolescent or smaller teenage child is to perform the entire colectomy via the Pfannenstiel incision with a wound protector for gentle retraction and optimal visualization.

FORMATION OF END ILEOSTOMY

The stoma is exteriorized at a previously marked site. A circle of skin about 2 cm in diameter is removed with cautery and the fascia is cruciated to allow the ileum to exit comfortably through it (**Figure 48.5a**). The TI is prepared for creation of the ileostomy by ligating a few vessels along the most distal 3 cm of small bowel. The ileum is tacked to the peritoneum with interrupted sutures (polyglycolic acid) (**Figure 48.5b**). Once the abdomen and skin are closed, a classic Brooke stoma is formed.



48.5a,b

Endorectal pull-through combined with double-stapled ileoanal anastomosis

POSITION OF PATIENT AND INCISION

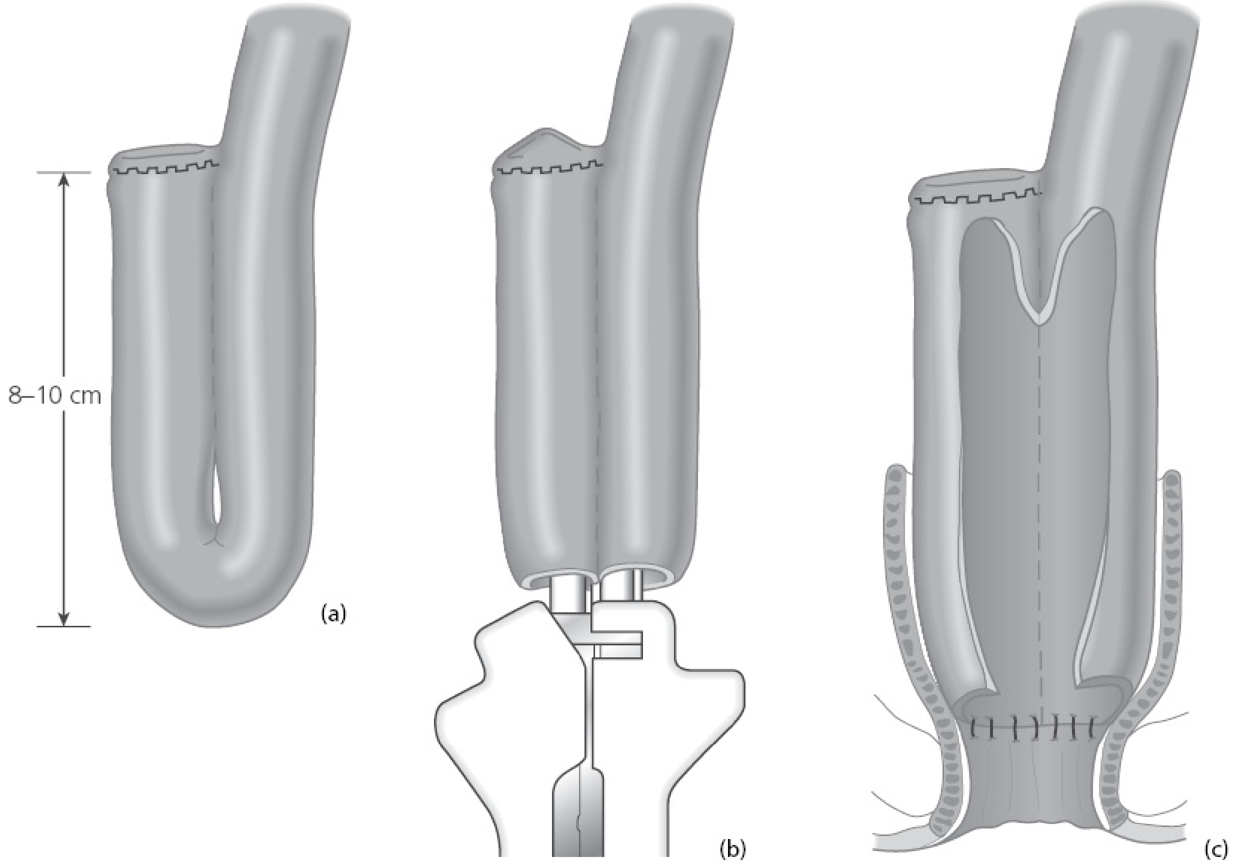
Positioning for this procedure is essentially identical to that for proctocolectomy. The legs can be prepped and draped and positioned so that the lower extremities fit properly in the stirrups in lithotomy; however, the stirrups are then repositioned so the legs do not impede the initial laparoscopic colon mobilization. Careful attention must be paid to padding of the lower extremities to avoid neurovascular injury. The incisions for port placement and abdominal colectomy proceed as already described.

MOBILIZATION OF ILEUM

The ileum is mobilized proximally to allow for an adequate length for the pull-through. In general, length is rarely a problem with a straight pull-through. With the J-pouch, more time must be spent in gaining bowel length. Adequate bowel length can also be difficult when the patient has had a previous ileostomy, as the mesentery may be scarred and foreshortened. In general, the main branch of the ileocecal artery can often be spared and the more proximal arcades ligated in the distal ileum. It is important to preserve the distal vascular arcade to the end of the ileum. If the ileocecal artery must be ligated, a bulldog clamp should initially be placed on this vessel to determine if a significant length of bowel will be lost. In such a case, one might opt for a straight pull-through, which will not require as much mobilization and length.

FORMATION OF RESERVOIR

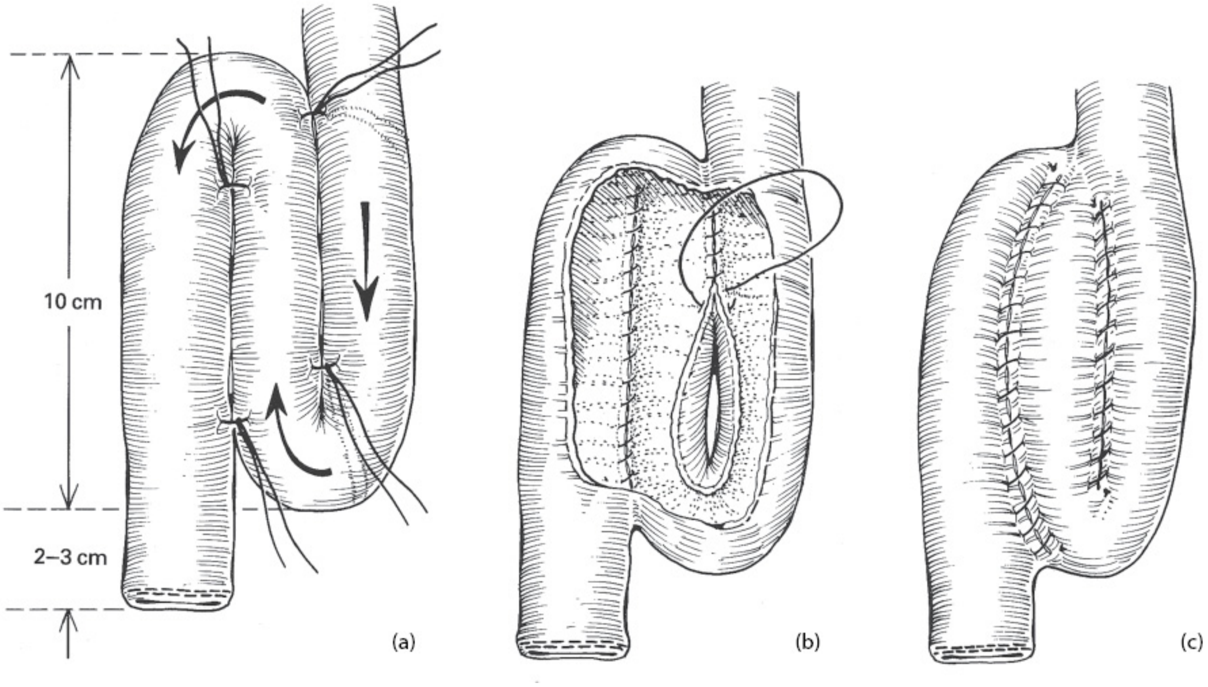
For the J-pouch, the ileum is folded back on itself for a length of 8–10 cm (**Figure 48.6a**). The bowel is opened with electrocautery at the apex and an Endo GIA□ stapler with white loads is fired to create the pouch (**Figure 48.6b**). A 3/0 silk suture is placed at the crotch of the staple line. Staple lines should be inspected for bleeding. It is critical that a septum is not left between the ileal limbs, as this will cause stasis, bacterial overgrowth, and a fecaloma. Next, the rectum is sized and the appropriately sized anvil for the circular stapling device is inserted into the enterotomy at the apex of the J-pouch and secured in place using a 3/0 Prolene purse-string suture (**Figure 48.6c**). Care is taken to ensure that the future end-to-end stapling does not impinge on the mesentery of the ileum.



48.6a–c

Although the J-pouch is the most popular of all the reservoirs, some surgeons use other pouches, such as the lateral ileal reservoir, the S-pouch, and the W-pouch. The W-pouch is rarely used in adults or children and is not discussed here.

The S-pouch is created in a very similar manner to the J-pouch, except that there are two overlapping limbs of ileum. Each limb is 10 cm long with a 2–3 cm spout, which is used for the ileoanal anastomosis (**Figure 48.7a–c**). The advantage of an S-pouch is an extra 1–2 cm of length and greater potential for a tension-free ileoanal anastomosis when a J-pouch is unable to reach. However, patients may develop efferent limb syndrome with partial obstruction of the pouch outlet.



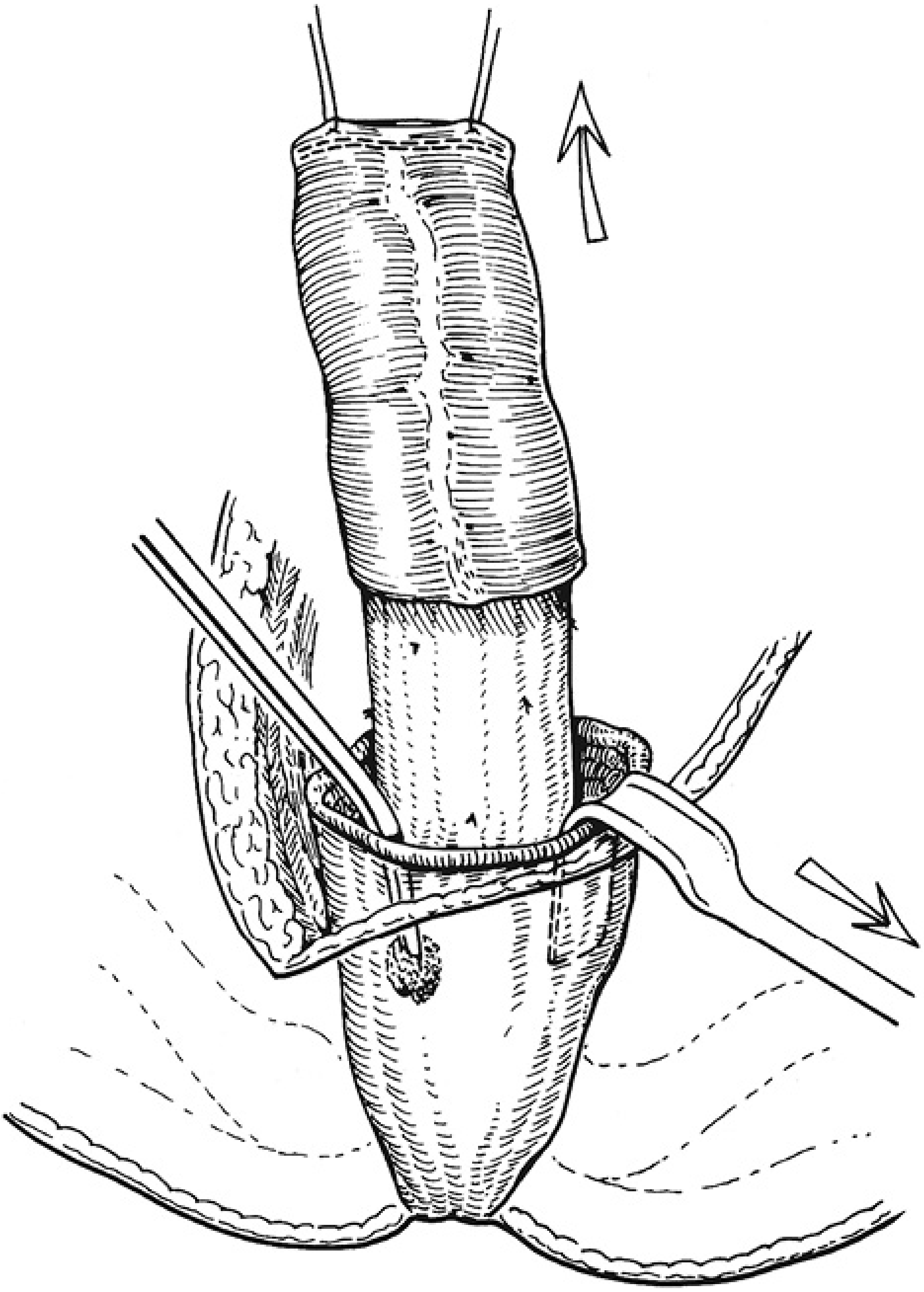
48.7a–c

ENDORECTAL DISSECTION

The authors prefer to do the entire endorectal dissection from the abdominal approach. It may be technically more difficult to develop the correct plane from the perineum, and, although the initial portion of the dissection can be quite difficult in children with UC, the dissection proceeds fairly smoothly once the correct plane is found.

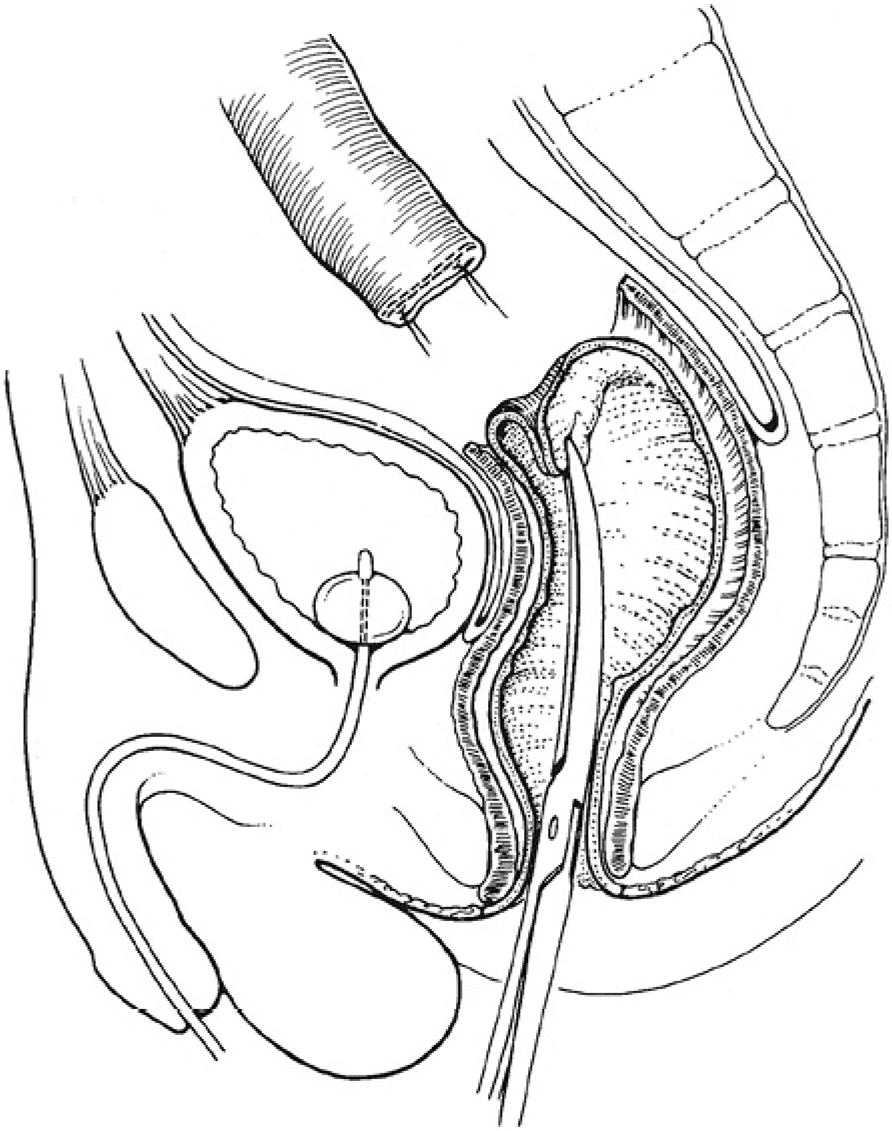
The endorectal mucosectomy is carried out from the level of the peritoneal reflection and extends down to about 1.5 cm from the dentate line. The peritoneal reflection is incised and the dissection is continued directly on the muscular rectal wall. The mesorectum is divided using the Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA) and Bovie electrocautery. The seromuscular wall of the rectum is divided and dissection is carried bluntly circumferentially down to the floor of the pelvis to the level of the levator muscles along the rectal submucosa. The level of the end of the dissection should be checked intermittently by feeling for an assistant's finger placed in the anus, just above the dentate line.

Countertraction of the dissected mucosal/submucosal tube, as well as traction in an upward and outward fashion on the muscular cuff (often with sutures retracted on all four quadrants), helps with the dissection. **Figure 48.8** depicts this step in the open approach.



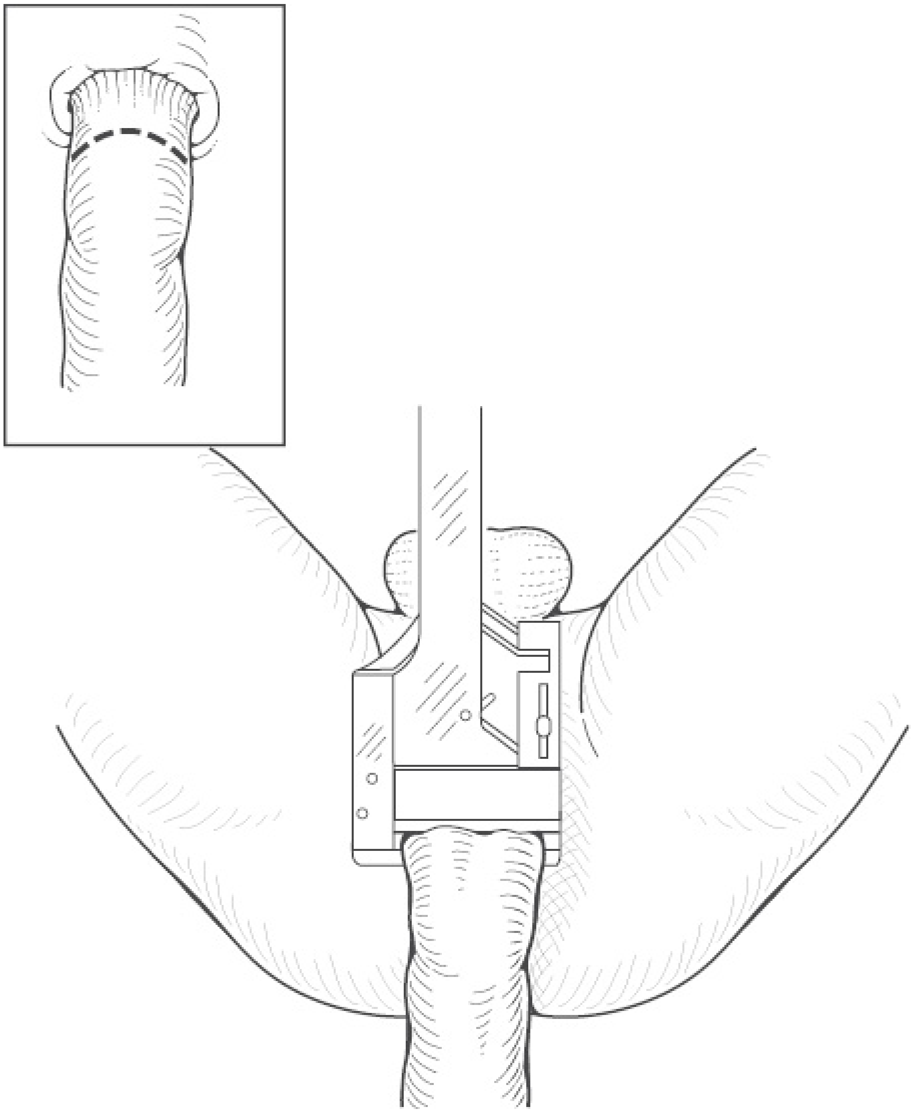
48.8

Once the endorectal dissection is completed and the rectum is circumferentially free, one of the surgeons moves to the foot of the table. Narrow retractors are placed at the anal mucocutaneous junction and a clamp is inserted into the rectum (**Figure 48.9**). An assistant working in the abdominal field places the end of the mucosal/submucosal tube into this clamp. The segment is then everted outside the perineum, taking care to maintain gentle traction on the muscular cuff intra-abdominally. The end of the everted tube is placed in a clamp and held on traction by an assistant.



48.9

The rectal stump is divided 1.5 cm above the dentate line with a fire of a transverse approximating stapler or Endo GIA□ stapler. In obese patients with a very deep gluteal cleft, the Endo GIA□-type stapling device allows better positioning of the stapler. The residual rectum is then inverted back into the pelvis (**Figure 48.10**) and the rectal specimen is sent to pathology.



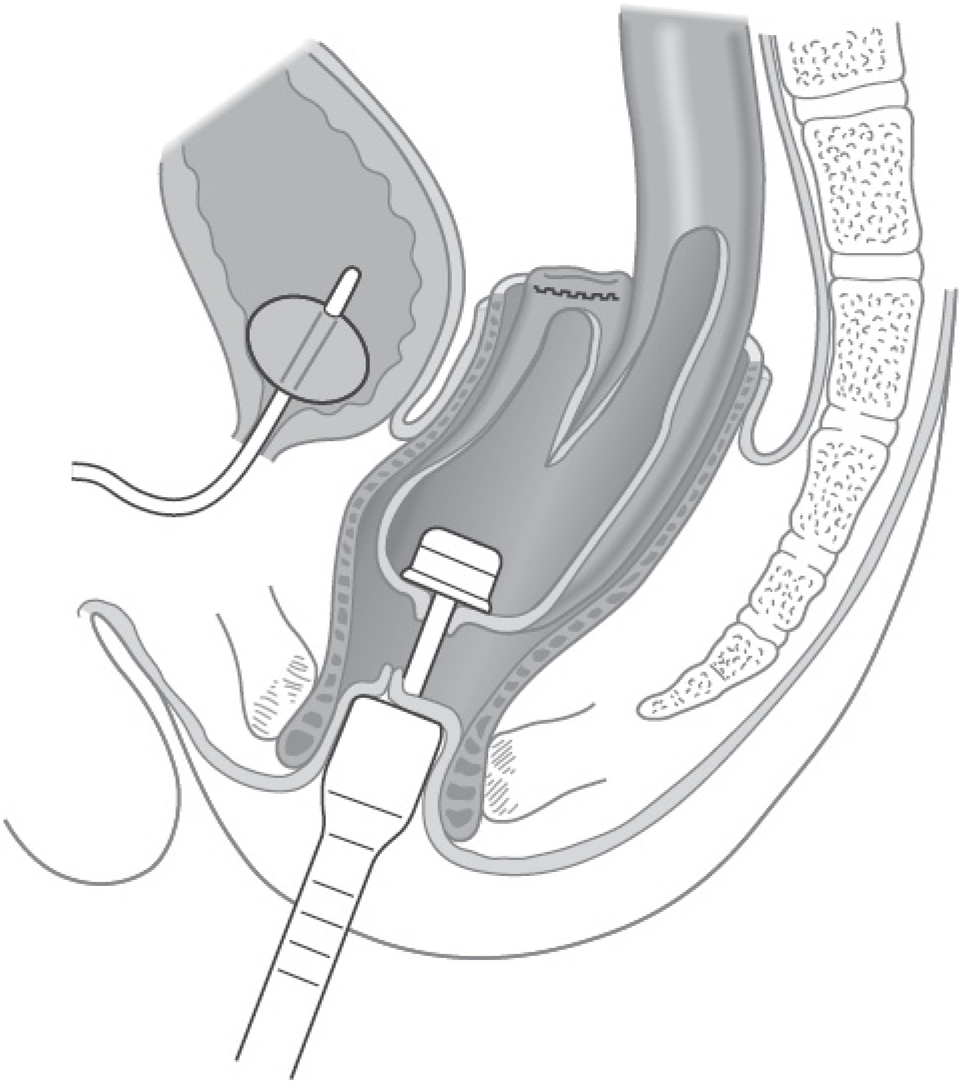
48.10

The dissection involves greater blood loss than in cases of Hirschsprung disease. Any large penetrating vessels should be cauterized, but bleeding usually stops spontaneously once the dissection is completed. If the correct plane is lost, the surgeon should turn to the opposite side of the bowel and work circumferentially until the dissection is distal to the area where the plane was lost.

DOUBLE-STAPLED ILEOANAL ANASTOMOSIS

The abdominal surgeon helps guide the J-pouch down toward the pelvis. At the same time, the surgeon on the perineal side turns the stapling device to fully advance the trocar in the mucosal/submucosal tube into the peritoneal cavity just posterior to the transverse staple line at the top of the anal canal. Great care must be taken to ensure correct alignment of the two sides, and to make sure both ends are cleared of adjacent tissues. Once this is done, the stapling device is fired and removed. Both tissue donuts are inspected and sent for pathology.

A flexible endoscope is gently inserted into the rectum to visualize the anastomosis (**Figure 48.11**). Air insufflation with the anastomosis submerged under irrigation fluid and the proximal bowel clamped between fingers is recommended to secondarily assess the integrity of the completed ileoanal anastomosis. The seromuscular cuff is sutured to the J-pouch circumferentially with interrupted 3/0 Vicryl□ suture. The J-pouch mesentery is sutured to the retroperitoneum with a running 4/0 PDS□ suture to prevent an internal hernia defect (**Figure 48.12**). A transanastomotic Penrose drain may be left in place to allow drainage of mucous for 2–3 postoperative days.



48.11

Loop ileostomy

The skin and fascia are incised as described previously like an end ileostomy, ensuring the fascia is cruciated to comfortably accommodate the loop ileostomy. A section of ileum appropriate for diverting ileostomy is identified and a 16 Fr red rubber catheter is passed through the mesentery at this level and used to bring up the ileum through the ostomy site with the proximal end cephalad and the distal end inferior.

POSTOPERATIVE CARE

Postoperative elements of ERAS include, among others, early enteral intake and mobilization, non-routine use of surgical drains and tubes, and limited use of narcotics. Implementation of isolated elements in the pediatric population have shown generally positive results, especially in the areas of mobilization, analgesia, selective use of drains and catheters, and early enteral feeding.[16]

After surgery, nasogastric decompression, if in place, may be removed on postoperative day 1, and enteral feedings can be advanced as tolerated. If the child has been malnourished, consideration should be given to continued postoperative parenteral nutrition. The Foley catheter in proctocolectomy and pull-through patients is usually removed on postoperative day 2 or day 3, but it may be left longer if the pelvic dissection was difficult. Patients in whom stress-dose steroids were administered should have their doses tapered over weeks. Initiation or continuation of other immunomodulatory or biologic therapy after surgery should be discussed with the gastrointestinal team and arranged prior to discharge if indicated. Children with new ostomies should receive routine stoma care and teaching for both patients and families.

COMPLICATIONS

Complications of surgical management of perianal disease in CD include recurrent abscesses and fistulas and can affect up to one-third of patients. After primary ileocecectomy for pediatric CD, complication rates, defined as any kind of morbidity within 30 days of surgery, have reported ranges of 30–45%. Severe complications (requiring surgical, endoscopic, or radiological intervention) occur in 10–27% and are usually secondary to wound complications and intra-abdominal abscess or anastomotic leak.[4]

Post-surgical complication rates have been found in 30–60% of all patients after IPAA.[17] Pouchitis is the most common complication affecting one-third to half of patients and usually treated with antibiotics. Randomized controlled trials in adults, albeit in small sample sizes, seem to demonstrate probiotics, specifically VSL 3 (a multibacterial culture for oral use), can be successful maintenance therapy for pouchitis, once remission is achieved.[18] The most serious complication, pelvic sepsis due to complete breakdown of the IPAA, was found in 0–13% in pediatric series,[19] and can often ultimately result in pouch failure. An anastomotic leak of any degree, occurring more often at the ileoanal anastomosis than from the pouch, also has a reported rate of 0–13% in pediatric series after RP.[20] Following the tenets of minimizing tension and maintaining good blood supply to anastomoses is crucial, and performing a “leak test” with air insufflation via flexible sigmoidoscope as described is common practice. Though a diverting ileostomy at the time of RP does not decrease the risk of developing an anastomotic leak, it may make management of a leak easier. Other management options, depending on the clinical scenario, include antibiotic therapy, transcutaneous drain placement by interventional radiology, creation of an end ileostomy or conversion of a diverting ileostomy to an end, and resection of the pouch itself. A novel technique for anastomotic leak gaining traction in adult colorectal surgery circles is application of the Endo-SPONGE® (B. Braun Medical Ltd, Sheffield, UK), or endoluminal vacuum-assisted therapy. A sponge connected to a vacuum on negative pressure placed into the presacral space endoscopically is exchanged every few days under sedation as the presacral cavity shrinks in size. Though the literature describes application in adults only thus far, and mostly of patients with benign colorectal disease or cancer, outcomes have been favorable for complete resolution of anastomotic leaks in shorter time compared to conventional methods.[21] One recent report describes application of the Endo-SPONGE® for anastomotic leak in eight patients after IPAA and resolution in all after a median of 60 days of therapy.[22] This technique is a promising adjunct for potential application in the pediatric IBD population.

Pouch failure rate is around 10% after IPAA in children, which is higher than that in adults (5–7%), and is attributed to potentially increased severity of disease in the pediatric population and associated negative effects on growth and development.[23] Ileoanal anastomotic stenosis is a common source of morbidity and found in 5–27% of children after RP.[19] They may be secondary to surgical technique (more often with stapled anastomoses than hand-sewn anastomoses), ischemia, concurrent use of NSAIDs, or features of CD. These usually are managed with serial dilations in the operating room. Patients who had a diverting ileostomy at the time of RP were more likely to present with anastomotic stenosis and require dilations than those without a protective ileostomy, suggesting that a defunctionalized pouch may contribute to development of the stenosis, either from excess fibrosis or inflammation.

Adhesive small bowel obstructions (SBOs) can occur after any intra-abdominal surgery, and in this case, postoperative patients with IBD do not differ. It is important to discriminate adhesive SBO from obstructive symptoms secondary to worsening stricturing disease in CD. Careful review of the patient's history (e.g. the patient's "typical" Crohn's symptomatology and phenotype) and contrasted cross-sectional imaging (e.g. magnetic resonance elastography, MRE), when appropriate, will be the most helpful.

Patients with IBD are at increased risk for *Clostridium difficile* infection (CDI), and different from adults, pediatric patients with UC and CD are equally susceptible to toxigenic *Clostridium difficile*-associated disease (CDAD) and asymptomatic colonization.[24] Children are also more likely to be asymptomatically colonized with *C. difficile*, so differentiating between an IBD flare with *C. difficile* colonization and CDAD is a challenge. Those with concomitant IBD and CDI are reported to have longer hospital stays and more often require parenteral nutrition and blood transfusions compared to pediatric patients with IBD alone. Escalation of IBD therapy has been found to be more rapid in children with a history of both IBD and CDI, though CDI was associated with a decreased risk of surgery and colectomy.

OUTCOME

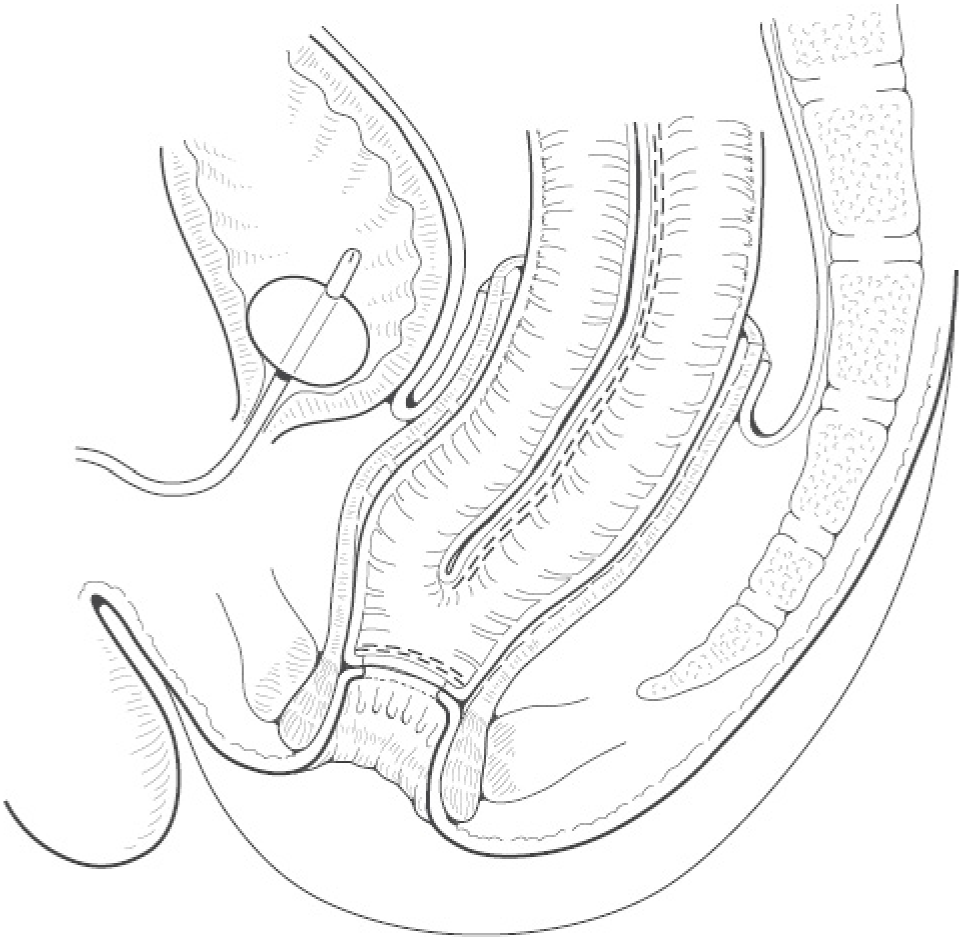
Clinical and surgical disease recurrence are the major concerning outcomes in pediatric CD and have been reported in 50–60% and 12–17% at 5 years, respectively.[4] Risk factors include female gender, microscopically positive resection margins, disease outside the ileocecal region, surgical resection, younger age at first surgery, and penetrating/stricturing phenotypes.[25] Adverse effects on surgical outcomes secondary to preoperative use and timing of anti-TNFα (tumor necrosis factor) agents is an ongoing debate, but several reports indicate immediate postoperative therapy may potentially postpone or prevent disease recurrence. Restoration of normal growth is a critical outcome in surgical intervention for CD as growth retardation is a significant cause for morbidity in children and indication for intestinal resection. Children have successfully demonstrated catch-up growth with improvement in growth rates after resection for CD during certain windows of growth potential, suggesting that surgery should be considered early as part of the armamentarium of therapy for CD and not just as a "last resort."

Clinical outcomes important to the pediatric patient with UC after RP include bowel movements per day (including night-time stools), urgency, and continence. Patients may have 7–15 loose bowel movements per day initially after reversal of the diverting ileostomy or after RP without ileostomy, and patients and families should be prepared and advised as such. Still, patients should be monitored for signs and symptoms of dehydration and/or evidence of skin breakdown by the perineum. Loperamide, Metamucil®, and occasionally cholestyramine may be prescribed to control the volume, frequency, and consistency of the stool. Over the course of several months, patients will settle on an average of 4–8 stools per day, with 0–1 per night. Younger patients tend to have more night-time bowel movements. Incontinence and seepage are rare, especially in long-term follow-up, and may be indicative of pouchitis; these symptoms and expectations are important to discuss with patients prior to surgery.

Colorectal cancer is of particular concern in patients with IBD and particularly with UC as described above. Literature in the adult population seems to show the prevalence and incidence of rectal or pouch carcinoma to be very low, and lowest after IPAA (<1%).[26] Risk factors include a diagnosis of UC (as opposed to CD), presence of residual rectum, duration of IBD disease, and presence of preoperative colorectal neoplasia. No specific guidelines exist for how often surveillance should occur in pediatric patients, but it is important to remind patients and families that residual rectal tissue is present even after RP and needs to be endoscopically evaluated.

ACKNOWLEDGMENTS

Figures 48.10–48.12 are reproduced from Geiger et al., *Surgery* 2003; 134: 492–5, with permission of the publishers, Elsevier, New York, NY, USA.



48.12

REFERENCES

1. Mulder DJ, Noble AJ, Justinich CJ, Duffin JM. A tale of two diseases: The history of inflammatory bowel disease. *J Crohns Colitis* 2014; 8: 341–8.
2. Crohn BB, Ginzburg L, Oppenheimer GD. Regional ileitis. A pathological and clinical entity. *JAMA* 1984; 251: 73–9.
3. Benchimol EI, Mack DR, Nguyen GC, et al. Incidence, outcomes, and health services burden of very early onset inflammatory bowel disease. *Gastroenterology* 2014; 147: 803–13.
4. Diederen K, De Ridder L, Van Rheenen P, et al. Complications and disease recurrence after primary ileocecal resection in pediatric Crohn's disease: A multicenter cohort analysis. *Inflamm Bowel Dis* 2017; 23: 272–82.
5. Adler J, Dong S, Eder SJ, et al. Perianal Crohn disease in a large multicenter pediatric collaborative. *J Pediatr Gastroenterol Nutr* 2017; 64: e117–e124.
6. Ceriati E, De Peppo F, Rivosecchi M. Role of surgery in pediatric ulcerative colitis. *Pediatr Surg Int* 2013; 29: 1231–41.
7. Kayton ML. Cancer and pediatric inflammatory bowel disease. *Semin Pediatr Surg* 2007; 16: 205–13.
8. Coran AG. A personal experience with 100 consecutive total colectomies and straight ileoanal endorectal pull-throughs for benign disease of the colon and rectum in children and adults. *Ann Surg* 1990; 212: 242–7.
9. Geiger JD, Teitelbaum DH, Hirschl RB, Coran AG. A new operative technique for restorative proctocolectomy: The endorectal pull-through combined with a double-stapled ileo-anal anastomosis. *Surgery* 2003; 134: 492–5.
10. Gray BW, Drongowski RA, Hirschl RB, Geiger JD. Restorative proctocolectomy without diverting ileostomy in children with ulcerative colitis. *J Pediatr Surg* 2012; 47: 204–8.
11. Hyams J, Markowitz J, Otley A, et al. Evaluation of the Pediatric Crohn Disease Activity Index: A prospective multicenter experience. *J Pediatr Gastroenterol Nutr* 2005; 41: 416–21.
12. Turner D, Hyams J, Markowitz J, et al. Appraisal of the Pediatric Ulcerative Colitis Activity Index (PUCAI). *Inflamm Bowel Dis* 2009; 15: 1218–23.
13. Rangel SJ, Islam S, St Peter SD, et al. Prevention of infectious complications after elective colorectal surgery in children: An American Pediatric Surgical Association Outcomes and Clinical Trials Committee comprehensive review. *J Pediatr Surg* 2015; 50: 192–200.
14. Nylund CM, Goudie A, Garza JM, et al. Venous thrombotic events in hospitalized children and adolescents with inflammatory bowel disease. *J Pediatr Gastroenterol Nutr* 2013; 56: 485–91.
15. Zitomersky NL, Levine AE, Atkinson BJ, et al. Risk factors, morbidity, and treatment of thrombosis in children and young adults with active inflammatory bowel disease. *J Pediatr Gastroenterol Nutr* 2013; 57: 343–7.
16. Shinnick JK, Short HL, Heiss KF, et al. Enhancing recovery in pediatric surgery: A review of the literature. *J Surg Res* 2016; 202: 165–76.
17. Alexander F. Complications of ileal pouch anal anastomosis. *Semin Pediatr Surg* 2007; 16: 200–4.
18. Ghouri YA, Richards DM, Rahimi EF, et al. Systematic review of randomized controlled trials of probiotics, prebiotics, and synbiotics in inflammatory bowel disease. *Clin Exp Gastroenterol* 2014; 7: 473–87.
19. Maruthachalam K, Bunn SK, Jaffray B. Complications following restorative proctocolectomy in children. *J Pediatr Surg* 2011; 46: 336–41.
20. Gonzalez DO, Nwomeh BC. Complications in children with ulcerative colitis undergoing ileal pouch–anal anastomosis. *Semin Pediatr Surg* 2017; 26: 384–90.
21. Strangio G, Zullo A, Ferrara EC, et al. Endo-sponge therapy for management of anastomotic leakages after colorectal surgery: A case series and review of literature. *Dig Liver Dis* 2015; 47: 465–9.

22. Rottoli M, Di Simone MP, Vallicelli C, et al. Endoluminal vacuum-assisted therapy as treatment for anastomotic leak after ileal pouch–anal anastomosis: A pilot study. *Tech Coloproctol* 2018; 22: 223–9.
23. Ozdemir Y, Kiran RP, Erem HH, et al. Functional outcomes and complications after restorative proctocolectomy and ileal pouch anal anastomosis in the pediatric population. *J Am Coll Surg* 2014; 218: 328–35.
24. Hourigan SK, Sears CL, Oliva-Hemker M. Clostridium difficile infection in pediatric inflammatory bowel disease. *Inflamm Bowel Dis* 2016; 22: 1020–5.
25. Splawski JB, Pffefferkorn MD, Schaefer ME, et al. NASPGHAN Clinical Report on Postoperative Recurrence in Pediatric Crohn Disease. *J Pediatr Gastroenterol Nutr* 2017; 65: 475–86.
26. Derikx L, Nissen LHC, Smits LJT, et al. Risk of neoplasia after colectomy in patients with inflammatory bowel disease: A systematic review and meta-analysis. *Clin Gastroenterol Hepatol* 2016; 14: 798–806.

49

Rectal polyps

JOSEPH L. LELLI JR

HISTORY

Juvenile polyps were first described by Verse in 1908 and for many years were considered adenomas and were treated with radical procedures. Horrilleno was the first to label the polyps that were found in children as juvenile polyps. However, it was not until 1962 that Morson demonstrated that juvenile polyps were benign hamartomas, distinguishing them from the potentially malignant adenomas. The histologic distinction between juvenile polyps and adenomas allowed for less radical interventions in children.

PRINCIPLES AND JUSTIFICATION

Rectal bleeding in young children is most commonly caused by anal fissures (>90%), with the next most common cause from a juvenile polyp. The presenting symptoms of polyps are bright red rectal bleeding (90%), with abdominal pain (10%) and prolapse through the anal canal (4%) making up other presenting symptoms.

Although the incidence of juvenile polyps is unknown, they are believed to occur in approximately 1% of all preschool children. Most appear in the first decade of life, with the peak incidence between 3 and 5 years of age. Polyps are solitary in 50% of cases, with the remainder having between two and ten polyps. Forty percent of juvenile polyps are found in the rectum or sigmoid colon. The remaining 60% are found evenly distributed throughout the colon.

Juvenile polyps are also known as retention, inflammatory, or cystic polyps. Such polyps are generally considered hamartomas or a malformation in which normal colonic tissue has become arranged in a haphazard manner. Grossly typical polyps have a glistening, smooth, spherical, reddish head and range from 2 mm to several centimeters in diameter. Polyps often have an ulcerated surface, which accounts for the rectal bleeding. Histologic cross-section demonstrates cystic spaces filled with mucus. Juvenile polyps are typically attached by a long, narrow stalk covered by colonic mucosa. The stalk predisposes the polyp to torsion, which results in venous congestion, abdominal pain, surface ulceration, bleeding, and not infrequently autoamputation.

PREOPERATIVE ASSESSMENT

Recent data suggest that juvenile polyps result from a structural rearrangement of the mucosa secondary to an inflammatory process. The initial event is probably ulceration and subsequent inflammation of the mucosa, leading to obstruction of regional, small colonic glands of the mucosa. The obstructed glands enlarge with mucous secretion and push up into the lumen. The fecal stream and peristalsis push the mass down the lumen, causing the stalk to elongate, resulting in the typical pedunculated appearance of the juvenile polyp. Ulceration of the surface or autoamputation leads to the bright red blood noted on presentation. Occasionally, polyps prolapse through the anal canal and present as dark, cherry-red protrusions at the anus. Although many of the polyps are within reach of a digital rectal examination, they may not be easy to feel due to their mobility. Clinical symptoms of intermittent bright red blood warrant an investigation.

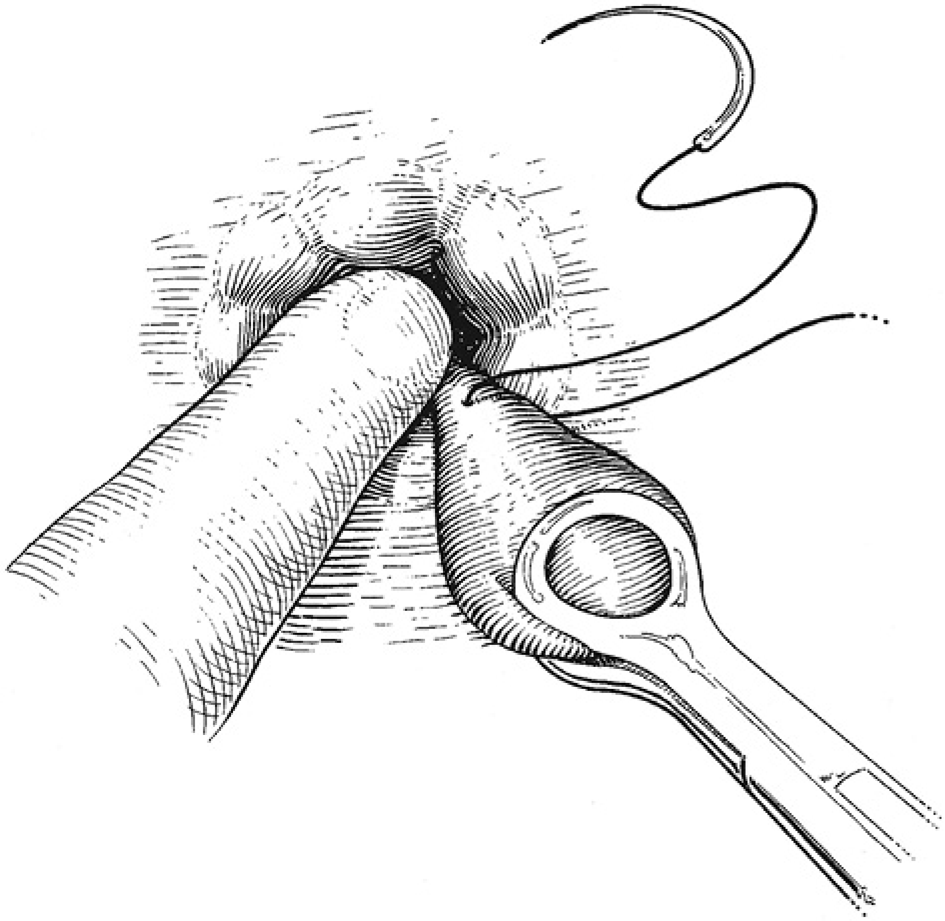
The diagnosis and treatment of juvenile polyps require a combination of history, digital rectal examination, and colonoscopy. The historic shift of juvenile polyps to the more proximal colon and the concern for the presence of juvenile polyposis (more than five juvenile polyps), with its increased risk of malignancy, mandates that the entire colon be surveyed. Polyps in the rectum can be removed easily during anoscopy. More proximal surveillance needs to be done by pancolonoscopy.

Differential diagnosis

The differential diagnosis of juvenile polyps encompasses all of the causes of rectal bleeding in toddlers and children up to the age of 6 years. Anal fissures and rectal prolapse cause rectal bleeding, but are easily distinguished from polyps on physical examination. Bleeding from a Meckel's diverticulum or duplication of the intestine usually causes more substantial blood loss than that from a polyp, and the blood usually mingles with the stool rather than coating it. Bleeding from an intussusception is accompanied by abdominal pain that is substantially worse than that seen with polyps. Inflammatory bowel disease is usually accompanied by diarrhea, which is not seen with polyps. Blood dyscrasias, such as Henoch–Schönlein purpura, should also be considered in the differential diagnosis. Rectal polyps occur in up to 15% of children with Peutz–Jeghers syndrome, and this condition should be considered.

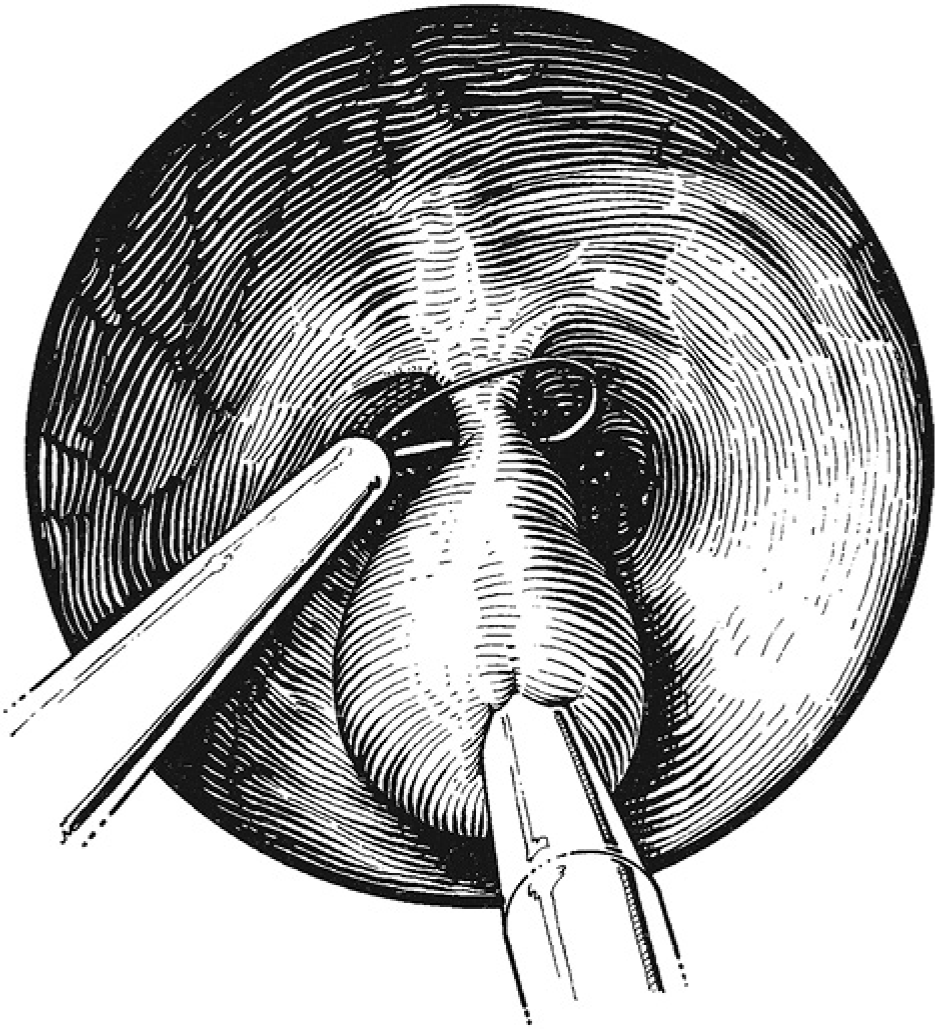
OPERATION

Anoscopy with polyp removal and pancolonoscopy can be performed as an outpatient procedure. Standard bowel preparation for a colonoscopy is performed by the patient's parents at home prior to surgery. Suitable premedication is given and general anesthesia is induced. The child is turned into the left lateral position with the sacrum at the edge of the operating table. The anus and perineum are inspected and digital rectal examination is performed. Polyps in the anorectal area can usually be removed at this point if they are found at the anal verge (**Figure 49.1**).



49.1

If a polyp is not identified on digital examination, an appropriately sized anoscope is lubricated and inserted. Since complete bowel preparation and cleansing have been performed, the presence of formed stool is unlikely. If stool is present, removal can be accomplished with a moistened cotton swab. When a polyp is identified deeper in the rectum, a chromic or Vicryl® ENDOLOOP® is placed around the base of the stalk and cinched down. A second ENDOLOOP® is placed at a distance distally on the stalk, tightened down, and the stalk cut. This allows for accurate placement of sutures and easy removal of the polyp once cut (**Figure 49.2**).



49.2

After all polyps have been removed within the area of the anoscope or if no polyps are found in the most distant rectum, pancolonoscopy is performed. All polyps are removed endoscopically by the standard snare technique. The location of each polyp is recorded.

A polyp in the lower rectum or at the anal canal verge can often be brought out through the anus by digital examination. The stalk is then ligated by an absorbable suture and then cut (see **Figure 49.1**). Often, the stalk is torsed and the blood supply thrombosed. These polyps are occasionally avulsed during the maneuver to bring them out of the anal canal. Avulsion may cause minor bleeding, which will usually stop spontaneously or by applying pressure with a cotton-tipped applicator. Persistent bleeding can usually be controlled by the application of electrocautery.

Polyps further up in the rectum can be visualized using an anoscope or nasal speculum placed in the anal canal. An absorbable ENDOLOOP® snare is looped over the polyp and tightened down at the mucosal base (see Figure 49.2). A second ENDOLOOP® is placed distally on the stalk and the stalk is then transected with scissors, electrocautery, or a scalpel. The polyp should be recovered for histologic examination, and this is easily accomplished by holding on to the distal ENDOLOOP® when the stalk is transected.

POSTOPERATIVE CARE

Most children can return home as soon as they have recovered from anesthesia. Parents should be warned that there may be some blood in the stool for a few days. The surgeon should follow up with the parents, verifying the type of polyp found on histologic examination. The parents should also be counseled to return should new bleeding occur.

COMPLICATIONS

Perforation and hemorrhage can occur, but they are extremely rare complications that are usually self-limiting.

OUTCOME

Most children with juvenile polyps will not have a recurrence of the problem. Pan colonoscopy should be performed, however, for any child with recurrent bleeding. In children with between two and four polyps, juvenile polyposis syndrome, with its increased risk of malignancy, should be considered. These children should undergo pancolonoscopy again a year after their initial polyp removal.

FURTHER READING

Franzin G, Zamboni G, Dina R, et al. Juvenile and inflammatory polyps of the colon: A histological and histochemical study. *Histopathology* 1983; 7: 719–28.

Jass JR. Juvenile polyposis. In: Phillips RKS, Spigelman AD, Thomson JPS (eds). *Familial Adenomatous Polyposis and Other Polyposis Syndromes.* Boston, MA: Little Brown, 1994.

Mestre JR. The changing pattern of juvenile polyps. *Am J Gastroenterol* 1986; 5: 312–14.

Morson BC. Some peculiarities in the histology of intestinal polyps. *Dis Colon Rectum* 1962; 5: 337–44.

Nelson EW, Rogers BM, Zawatsky L. Endoscopic appearance of auto-amputated polyps in juvenile polyposis coli. *J Pediatr Surg* 1977; 12: 773–6.

50

Acquired anorectal disorders: Prolapse, fistula, and hemorrhoids

MICHAEL STANTON

INTRODUCTION

Acquired anorectal disorders in children include anal fissures, perianal abscess/fistula, hemorrhoids, and rectal prolapse. A digital photograph taken by the child's parents is often very helpful in making the diagnosis. In general, fissures, hemorrhoids, and rectal prolapse are initially treated by attempting to normalize stooling. This is by dietary modification, stool softeners (e.g. Laxido, Movicol) and/or stimulant laxatives (e.g. senna). If medical management fails, then surgical interventions may be indicated.

RECTAL PROLAPSE

PRINCIPLES AND JUSTIFICATION

Although rectal prolapse is common and usually resolves spontaneously, in a proportion symptoms will persist despite normalization of stooling. Mucosal prolapse is described in infants, whereas adolescents are likely to have full-thickness prolapse (as seen in adults). A proposed mechanism is of stretching to the levator mechanism provoked by either constipation or diarrhea. The typical circumferential rosette of rectal mucosa is seen at the anal verge after stooling. The child may be aware of the sensation of a swelling. There may be associated bleeding, urgency, pain or tenesmus. The child may be able to reduce prolapse by gluteal/perineal muscle contraction. Manual reduction by the parents (or rarely as an emergency for irreducible prolapse) may be required. Only a minority of infants will require injection sclerotherapy. Rectopexy may be indicated in adolescents with ongoing symptoms.

PREOPERATIVE INVESTIGATIONS

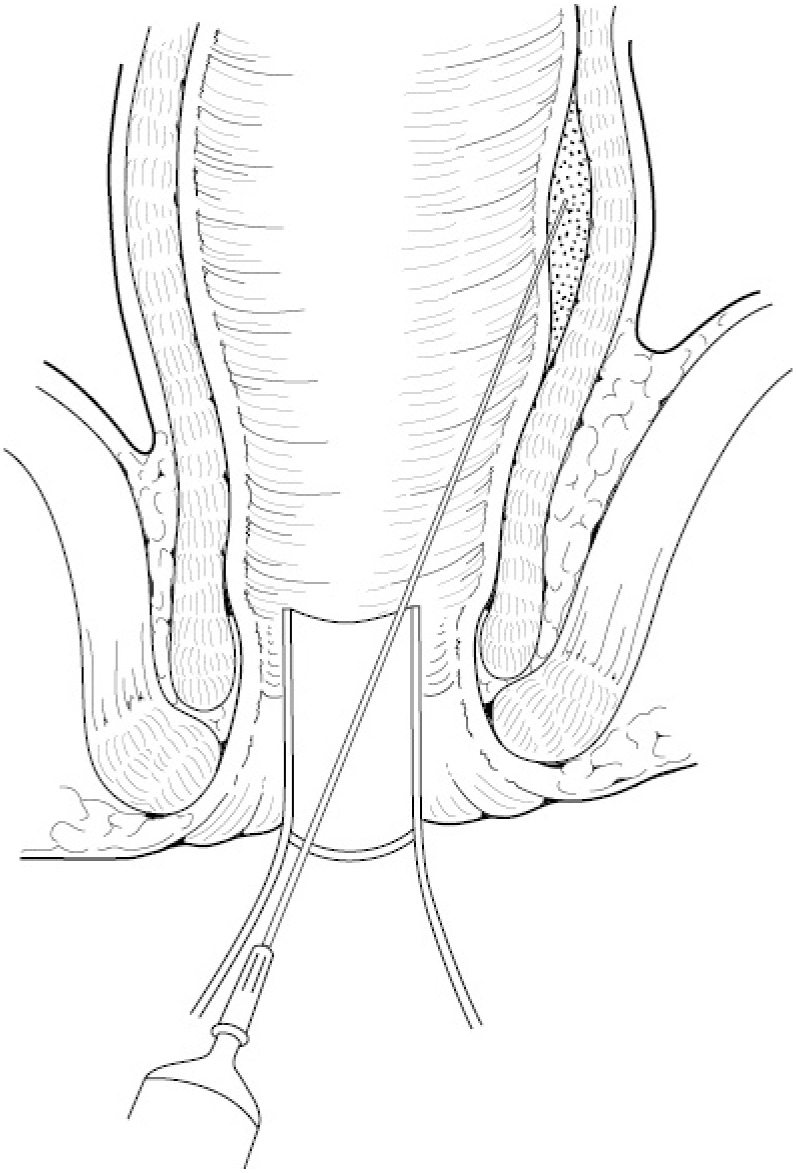
The diagnosis is usually obvious from the history and a digital photograph taken by the parents. An underlying diagnosis such as cystic fibrosis should be considered in chronic prolapse with failure to thrive though since the introduction of newborn bloodspot screening (in 2007 in the UK, for example), this is now a rare presentation.

OPERATIONS

Injection sclerotherapy

For infants, injection sclerotherapy of the non-adherent rectal mucosa is undertaken under general anesthesia. Oily phenol (5%) or hypertonic saline (30%) are commonly used.

The lithotomy position is used. Rigid sigmoidoscopy is performed to exclude other diagnoses; rectal polyp may mimic prolapse. A Park's or Killian's retractor is placed in the anal canal. Submucosal injection above the dentate line is performed in three positions (away from the posterior midline), raising a visible bleb (**Figure 50.1**).



50.1

Recurrence may occur in up to one-third of cases, necessitating repeat injection.

Laparoscopic abdominal rectopexy

Rectopexy is seldom performed as an open procedure now given the technological advances that have been made. As in adult practice, there is a debate as to whether non-absorbable mesh should be used to reinforce the repair in children.

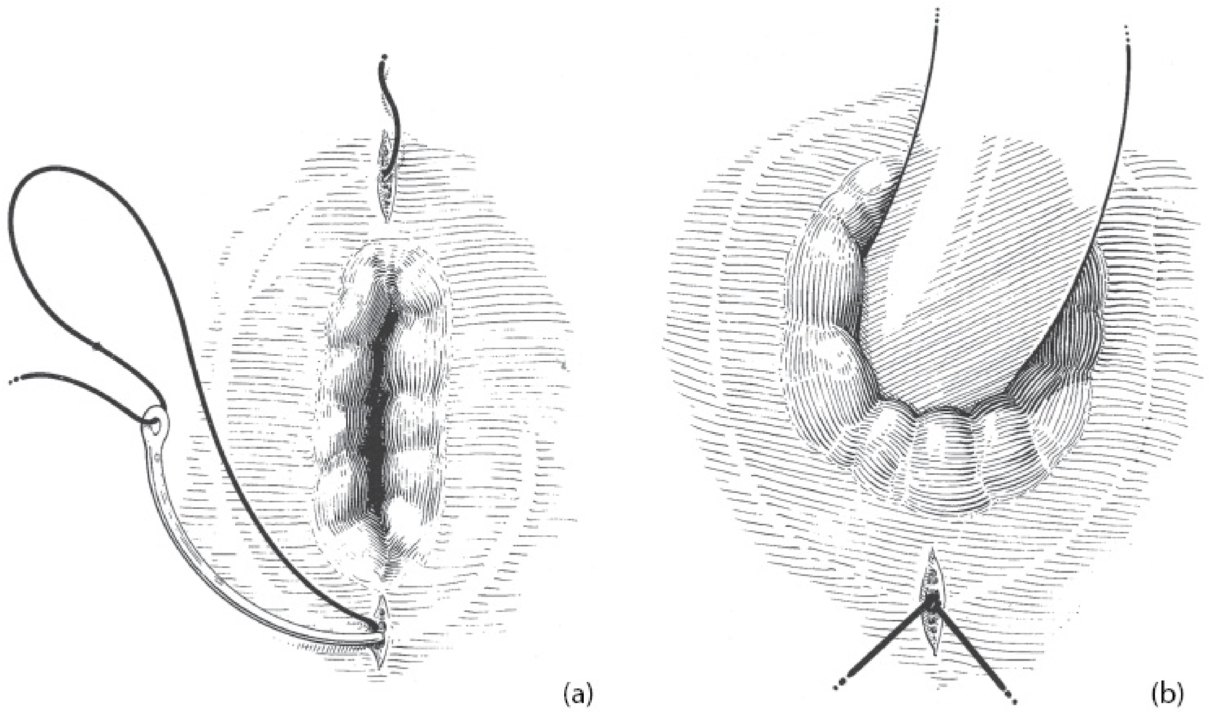
Trendelenburg position is used with three- or four-port laparoscopy.

The rectum is mobilized anteriorly and laterally. Care is taken to avoid pelvic nerves and ureters by dissecting close to the rectal wall. The redundant rectum is elevated out of the pelvis. The peritoneum overlying the sacral promontory is opened down to the level of the coccyx. The posterior rectal wall is sutured to the sacral promontory. Non-absorbable mesh may reduce recurrence rates. Rectosigmoid resection is undertaken by some surgeons to avoid postoperative constipation.

Perineal repair

Pelvic nerve damage may occur following abdominal rectopexy, leading to, for example, impotence in males, and more direct perineal approaches have been advocated. These include rectosigmoidectomy (Altemeier procedure), encirclement suture (Thiersch) or rectal mucosal excision (Delorme's procedure). Although these procedures reduce the risk of pelvic nerve damage, recurrence rates are higher.

In the Thiersch procedure two small incisions are made at 6 o'clock and 12 o'clock (**Figure 50.2 and b**). A length of absorbable suture (e.g. 2/0 PDS□) is threaded around the anus, deep to the external sphincter, and secured in place around a finger or Hegar dilator. The idea is to inhibit sphincter dilatation thus preventing prolapse.



50.2a,b

COMPLICATIONS/OUTCOME

Short-term recurrence of up to 5% is reported in pediatric series after surgical repair. Longer-term studies in adults and children show that this may rise to 20% or more after 10 years. Constipation is likely to continue or even be worse, and medical therapy should be continued.

ANAL FISSURE

HISTORY

The first description of anal fissure was in 1825 (in Boyer's *Traite des maladies chirurgicales*), where posterior sphincterotomy was described as a treatment. Soon after, in 1829, Recamier proposed anal dilatation to treat fissures. Various treatments have been used in children and in all cases a balance of recurrence rates versus complications (principally damage to the sphincter mechanism) must be made. In any case, most will respond to medical therapy alone.

PRINCIPLES AND JUSTIFICATION

Anal fissure is a longitudinal split in the anal margin, often occurring in the anterior or posterior midline position. The peak incidence is in children around 2 years of age. Fissure is usually provoked by passage of a large, hard, painful stool. Symptoms are of acute, severe pain on defecation, often with bright red rectal bleeding. Stool-withholding behavior may result in anal fissure in younger infants. The fissure may be seen in physical examination. An anterior skin tag ("sentinel pile") may be seen in chronic cases.

Medical treatment consists of stool softeners and stimulant laxatives. For refractory cases, more specific therapy may be used aimed at reducing anal sphincter resting pressure (which is thought to cause ischemia and poor healing). Glyceryl trinitrate ointment (e.g. 2%) has been used by some authors, but may be no more effective than intensive bowel management alone. Intrasphincteric botulinum toxin injection reduces resting sphincter pressure via cholinergic neuromuscular blockade. As the effects are reversible and there is no risk of long-term sphincter disruption, this has become the commonest procedure undertaken in children.

PREOPERATIVE INVESTIGATIONS

Further investigation is usually not required. Physical abuse should be considered in atypical cases (e.g. multiple radial fissures).

OPERATIONS

Botulinum toxin injection

Four quadrant injection of Botulinum toxin (e.g. Dysport®) is the author's preference. The maximum dose is 400 i.u., equating to 100 i.u. per quadrant. Confirmation of the sphincter position can be performed either with per rectal digital palpation or concomitant endoanal ultrasound. The typical percutaneous injection site is approximately 1 cm from the anal verge with the needle advanced by approximately 1–2 cm to the sphincter mechanism.

Other surgical procedures

Controversy still exists as to whether other surgical procedures should be carried out in infants and children. It is generally accepted that digital anal dilatation is associated with poor fissure resolution and is uncontrolled and so leads to higher rates of incontinence.

FISSURECTOMY

Fissurectomy has been described in children, and avoids the risk of sphincter damage. Stay sutures are placed either side of the affected area to demonstrate the fissure, which is then excised with full-thickness dissection of the anal mucosa. The mucosal defect is closed with absorbable sutures.

LATERAL SPHINCTEROTOMY

Although lateral sphincterotomy produces good healing rates, it is associated with a high incontinence rate and so has fallen completely out of favor in pediatric practice.

PERIANAL ABSCESS AND FISTULA

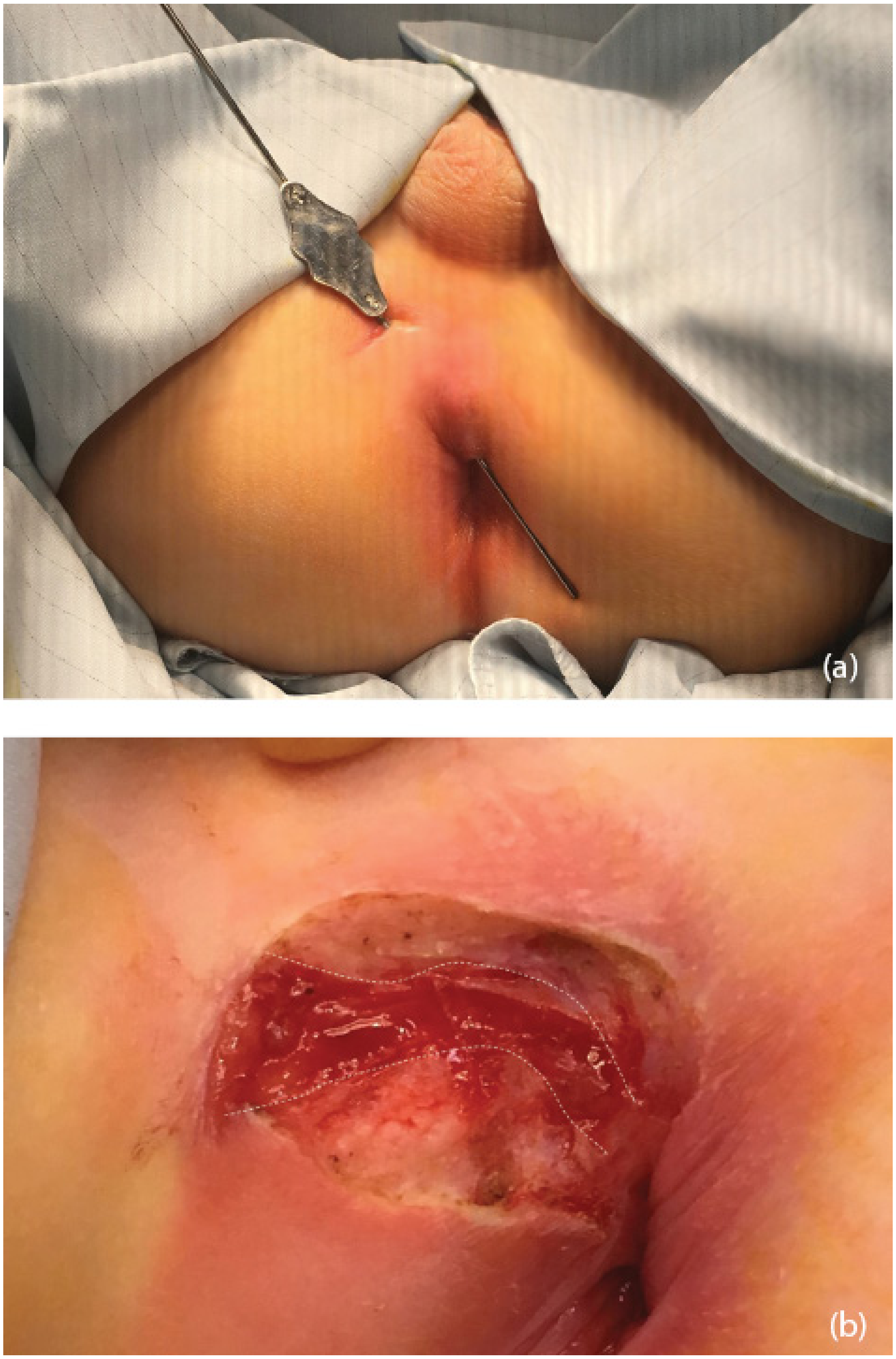
PRINCIPLES AND JUSTIFICATION

Perianal abscess is common in young infants (<2 years) with a further peak in teenagers. Crohn's disease should be considered in older children and a skin biopsy taken at the time of abscess drainage. It has been proposed that anal cryptoglandular abnormalities may lead to abscess formation. Abscess aspiration may be undertaken in infants (with prior topical local anesthetic). Incision and drainage are otherwise required for a fluctuant abscess. Subsequent fistula formation may occur in up to 20% of cases, although it remains controversial as to whether probing for a potential fistula track should be undertaken at the time of initial abscess drainage. Most authors perform drainage alone, due to the risk of possible tissue damage.

Anal fistula is a communication between the perianal skin and the anal canal following perianal abscess. Fistulas are "low" in infants/children and usually pass straight between the skin sinus and the anal canal at the level of the dentate line. Presentation is of a perianal abscess treated either surgically or, more often, incompletely by antibiotics alone, resulting in a chronically discharging perianal sinus. "Flare-ups" of recurrent painful swelling and redness may occur.

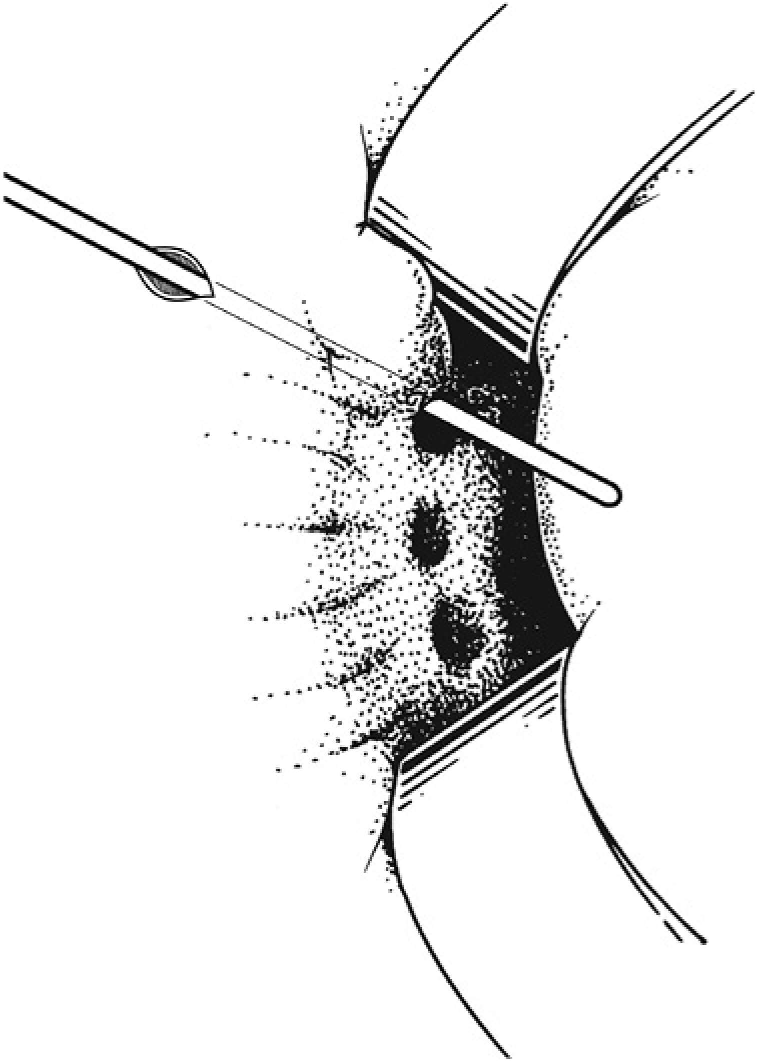
OPERATIONS

General anesthesia and lithotomy position are used. A nasal speculum is inserted to display the potential fistula track. A lacrimal or Lockhart–Mummery probe is passed along the fistula from the perianal opening into the anal canal (**Figure 50.3a**). Fistulotomy consists of direct incision of the perianal skin and subcutaneous onto the probe, using monopolar needlepoint diathermy. The base of the track may be carefully curetted if there is significant granulation tissue (**Figure 50.3b**). No dressing is required.



50.3a,b (a) “Lacrimal duct probe” outlining congenital fistula at 10 o’clock; (b) fistula laid open with mucosal track easily identified (and subsequently excised). (Images courtesy of Mark Davenport.)

In an older child where Crohn's disease is suspected, placement of a loose, non-cutting seton across the fistula may be safer while other investigations are carried out. A Park's retractor is used to display the anatomy. A probe (Lockhart–Mummery or probe with an eyelet) is passed along the fistula (**Figure 50.4**). Either a vascular sling or a braided non-absorbable suture is looped around the fistula and tied loosely. Skin biopsy is recommended.



50.4

HEMORRHOIDS

PRINCIPLES AND JUSTIFICATION

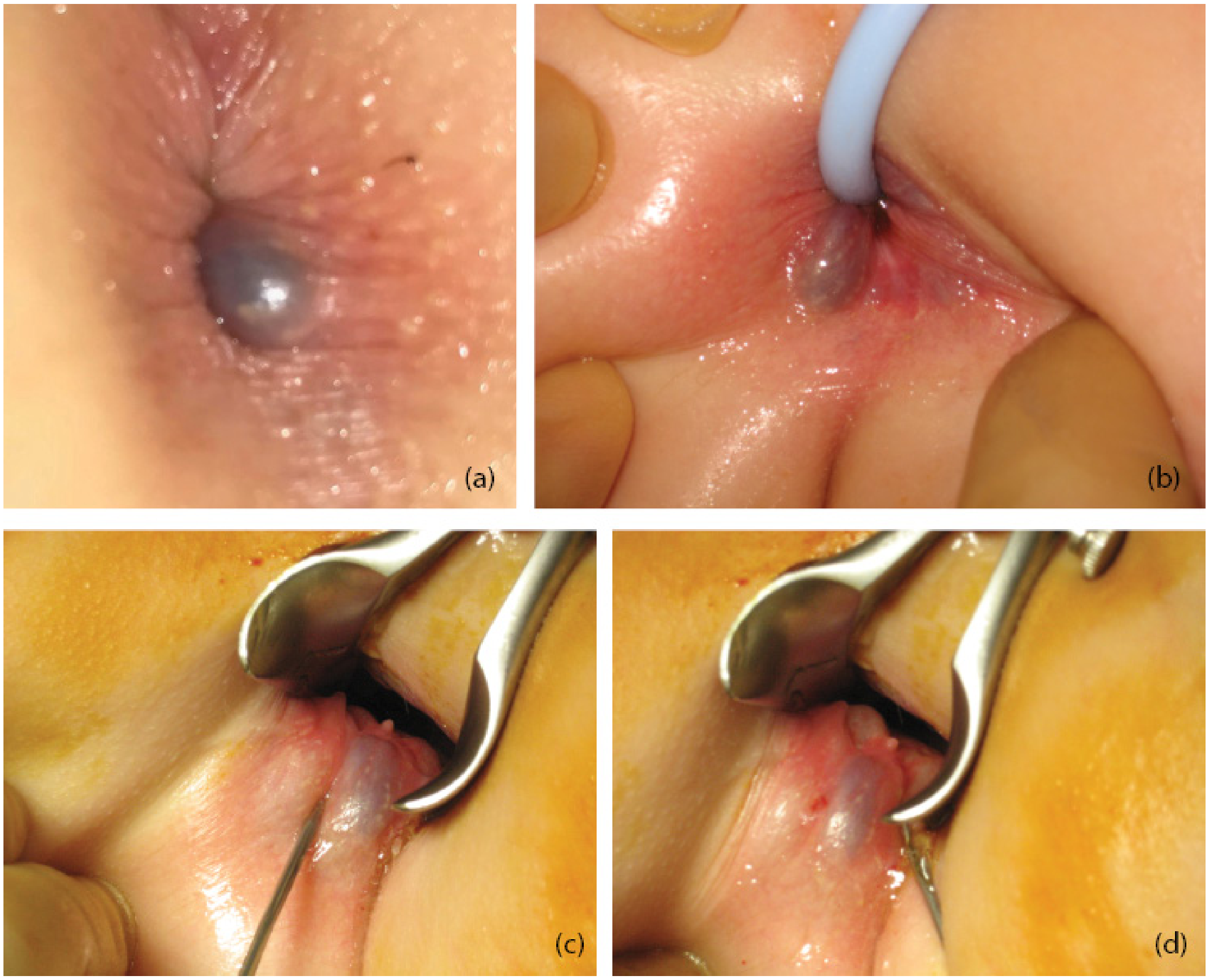
Hemorrhoids are relatively unusual in children/adolescents. Rarely they may present in association with anorectal varices in those with portal hypertension. These, however, are rarely *de novo* events with virtually all of these having had treatment of upper gastrointestinal varices previously.

Presentation is with a perianal swelling, with bleeding, though itching and tenderness are also possible. This is often preceded by constipation with straining. The swelling is usually a small bluish/purple lump at the anal margin, rather than the more obvious prolapsing swellings seen in adults. The swelling usually corresponds to the typical sites of the anal cushions (3 o'clock, 7 o'clock, and 11 o'clock).

Medical treatment including reassurance and management of constipation is often enough.

OPERATION

Surgery for hemorrhoids is required if conservative management is unsuccessful (**Figure 50.5a–d**). Rubber band ligation is not suitable in children. A large balloon catheter is passed into the rectum while the patient is in the lithotomy position, and withdrawn through the anal canal to demonstrate the hemorrhoid(s). Injection sclerotherapy (5% oily phenol) adjacent to the hemorrhoid is undertaken. Local anesthetic (and/or caudal anesthesia) should be used if the injection is close to the dentate line.



50.5a–d (a) Hemorrhoid; (b) insertion of balloon catheter; (c) parahemorrhoidal injection; (d) final result.

Hemorrhoidectomy (Milligan–Morgan, stapled or using a vessel-sealing device such as a LigaSure [Valleylab, Boulder, CO, USA]) is very rarely required in pediatric practice.

FURTHER READING

Babu R, Burge DM. A technique to demonstrate external haemorrhoids. *Pediatr Surg Int* 2004; 20: 476–8.

EL-Chammas KI, Rumman N, Goh VL, et al. Rectal prolapse and cystic fibrosis. *J Pediatr Gastroenterol Nutr* 2015; 60: 110–12.

Foppa C, Martinek L, Arnaud JP, Bergamaschi R. Ten-year follow up after laparoscopic suture rectopexy for full-thickness rectal prolapse. *Colorectal Dis* 2014; 16: 809–14.

Grossmann O, Soccorso G, Murthi G. LigaSure hemorrhoidectomy for symptomatic hemorrhoids: First pediatric experience. *Eur J Pediatr Surg* 2015; 25: 377–80.

Heaton ND, Davenport M, Howard ER. Incidence of haemorrhoids and anorectal varices in children with portal hypertension. *Br J Surg* 1993; 80: 616–18.

Husberg B, Malmborg P, Strigård K. Treatment with botulinum toxin in children with chronic anal fissure. *Eur J Pediatr Surg* 2009; 19: 290–2.

Kenny SE, Irvine T, Driver CP, et al. Double blind randomised controlled trial of topical glyceryl trinitrate in anal fissure. *Arch Dis Child* 2001; 85: 404–7.

Lambe GF, Driver CP, Morton S, Turnock RR. Fissurectomy as a treatment for anal fissures in children. *Ann R Coll Surg Engl* 2000; 82: 254–7.

Randall J, Gallagher H, Jaffray B. Laparoscopic rectopexy for external prolapse in children. *J Pediatr Surg* 2014; 49: 1413–15.

Sun C, Hull T, Ozuner G. Risk factors and clinical characteristics of rectal prolapse in young patients. *J Visc Surg* 2014; 151: 425–9.

51

Colostomy formation and closure

YEW-WEI TAN AND JOSEPH I. CURRY

HISTORY

Alexis Littre (1658–1726), the Parisian anatomist, is credited with being the first to propose a planned colostomy, or artificial anus. This occurred in 1710 during an autopsy on a child with anal atresia, when he explained how, after making an incision in the abdomen, it would be possible "to bring the superior part of the bowel to the abdominal wound, which should never be closed and which would perform the function of the anus." It was not until 1776 that this concept was applied, when a colostomy was performed by Pillore of Rouen on an adult with rectal carcinoma.

In 1783, Dubois is said to have been the first to construct a colostomy in an infant, but the patient, who had anal atresia, died after 10 days. The first long-term survivor was an infant, also with imperforate anus, who had a colostomy made by Duret in 1793 and was still alive 45 years later. During the 19th century, the procedure was introduced at centers across Europe and various modifications were developed, including loop colostomy over a rod by Maydl (1888). Operation *"a deux temps"* reported in 1885 by Davies-Colley of Guy's Hospital, London, consisted of suturing the bowel to the skin followed by delayed opening once the wound edges had sealed; this was an important development in an era when infection was the major cause of morbidity. The Hartmann procedure, described in 1923 for use following rectal excision for carcinoma at a time when anastomosis of the colon to the rectum was still dangerous, is still widely used in pediatric surgery.

Colostomy closure, too, was a dangerous procedure with a high risk of leakage and fecal peritonitis. This led to the introduction of techniques to minimize the risk of peritoneal contamination, such as extraperitoneal closure. With the Mikulicz technique, the common walls of a double-barreled colostomy are crushed using an enterotome or forceps, and the resulting fecal fistula is closed later, leaving the suture line extraperitoneal if desired. Today, colostomy formation and closure are safe operations provided basic surgical principles are adhered to, but the risk of potentially serious complications is ever-present and must not be underestimated.

PRINCIPLES AND JUSTIFICATION

A colostomy is used to divert the fecal stream from the distal colon and rectum, and may be temporary or permanent. A temporary colostomy may be a primary procedure in the management of a congenital anorectal anomaly such as Hirschsprung disease, following an injury, or to treat severe inflammatory bowel disease or infection. It may also be a secondary procedure to protect a distal anastomosis. A permanent colostomy is required following radical excision of the anorectum for disease or debilitating fecal incontinence, for example due to anal sphincter dysfunction or colorectal dysmotility. The following types of colostomy are in common use:

- *Divided colostomy.* This has the advantage of complete fecal diversion and a low risk of prolapse or retraction. The colon is transected and the distal end exteriorized as a mucous fistula and either placed adjacent to the proximal stoma so that both stomas may be enclosed within the same colostomy bag, or the stomas are widely separated by placing them at opposite corners of the incision so that the proximal, but not the distal, stoma lies within the bag in order to prevent feces entering the distal colon. Alternatively, the distal stoma is closed and replaced within the abdominal cavity (Hartmann procedure) (**Figure 51.10**).

- *Single-end colostomy.* In some circumstances, the colostomy is likely to be required for a longer duration or to be permanent. In these circumstances there is an advantage in making an end colostomy through a custom-made circular incision to offer the best potential for application of a stoma bag (without a scar going into the colostomy), and the least chance of stoma prolapse and parastomal hernias.
- *Loop colostomy.* A loop of colon is exteriorized and opened, but not divided. The procedure has the advantages of being simple and quick to perform and is of particular value in seriously ill patients, but it carries a higher risk of prolapse and retraction. The extent of fecal diversion is variable, which is a disadvantage in the presence of a complete anal occlusion, when the rectum cannot be washed out, or with an anorectal fistula, where there is a risk of recurrent urinary tract infection from rectal organisms.

Siting the colostomy

An elective colostomy is usually sited either in the proximal transverse or in the proximal sigmoid colon, where the colon is readily accessible and sufficiently mobile to be exteriorized without tension. The sigmoid colostomy has the advantage that there is a greater length of proximal colon available for fluid and electrolyte absorption, and the stools are thicker.

Optimal placement of the colostomy is important and will depend on the clinical situation. The colostomy bag must fit comfortably over the proximal stoma and, to avoid leakage, the surrounding skin surface must be flat, avoiding bony prominences, to ensure watertight adherence of the bag. Patients who manage their own colostomy, particularly those who are wheelchair-bound, must be able to access the stoma easily; this requires preoperative planning with a stoma therapist.

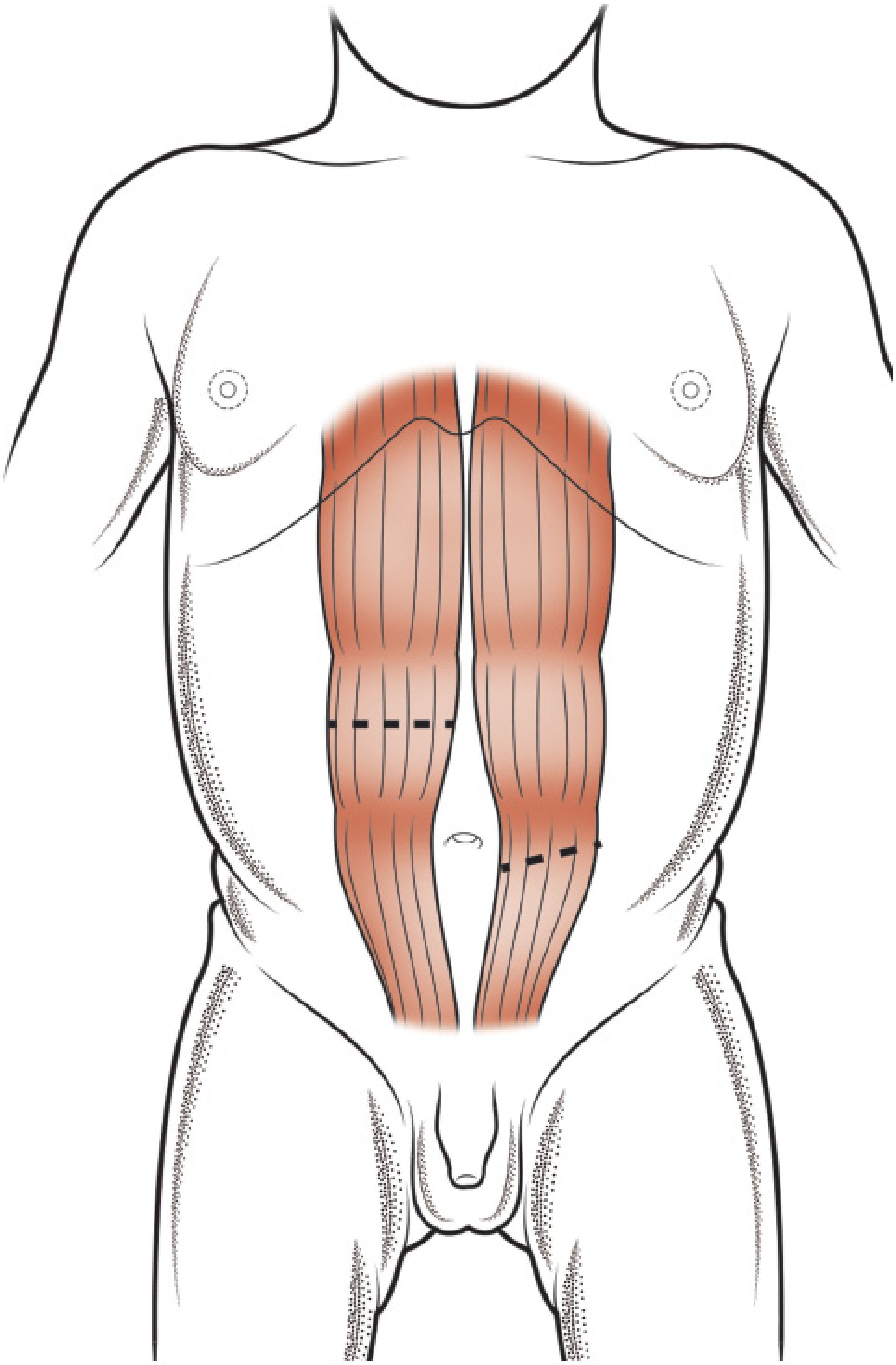
PREOPERATIVE MANAGEMENT

Colostomy formation is a major operation and the patient's condition must be optimized before operation. Nasogastric drainage and IV fluid therapy are indicated in the presence of obstruction. Prophylactic antibiotics are given preoperatively.

OPERATIONS

Divided colostomy

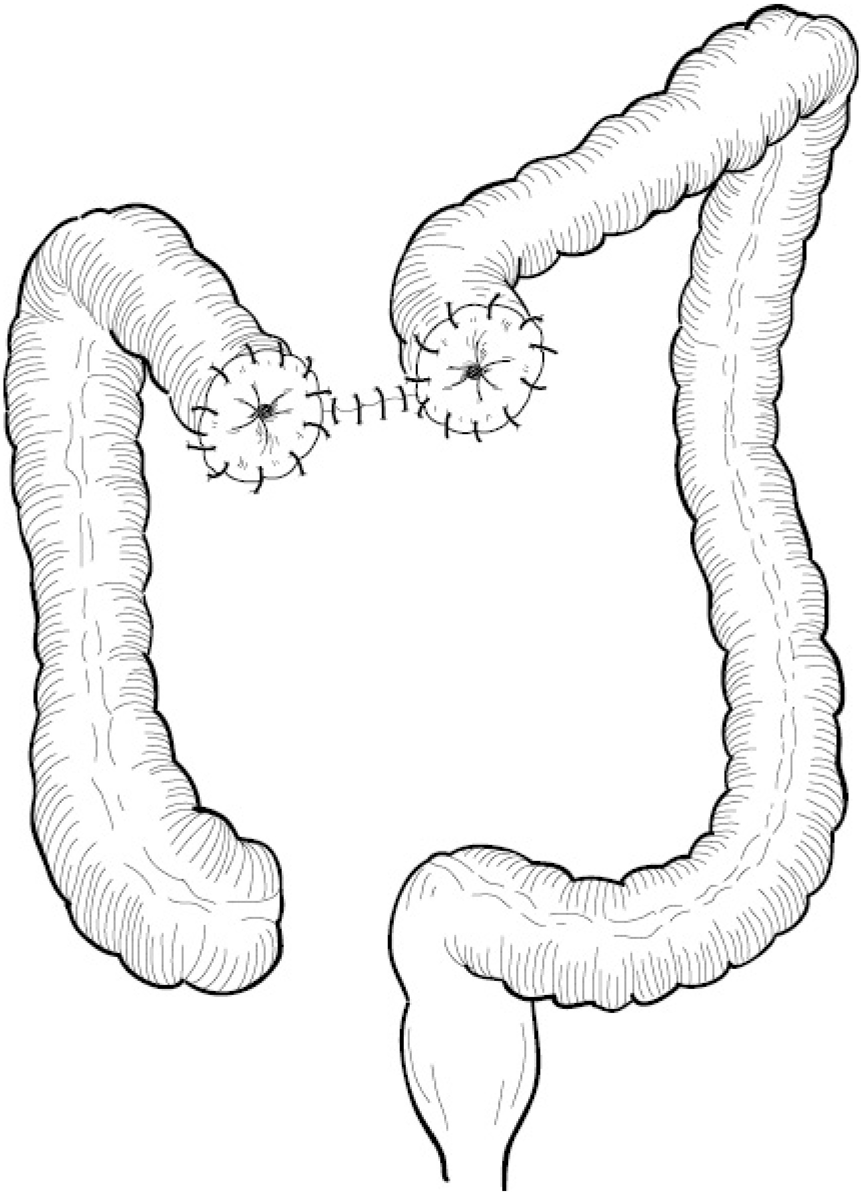
A right upper transverse incision is used for a transverse colostomy; a transverse muscle-sparing incision is used for a sigmoid colostomy, centered about midway between the umbilicus and the left anterior superior iliac spine (**Figure 51.1**). These positions are chosen for their proximity to the natural peritoneal fixation point for the hepatic flexure and the descending colon respectively so that a stoma formed just distal to these areas should potentially limit the extent of possible bowel prolapse.



51.1

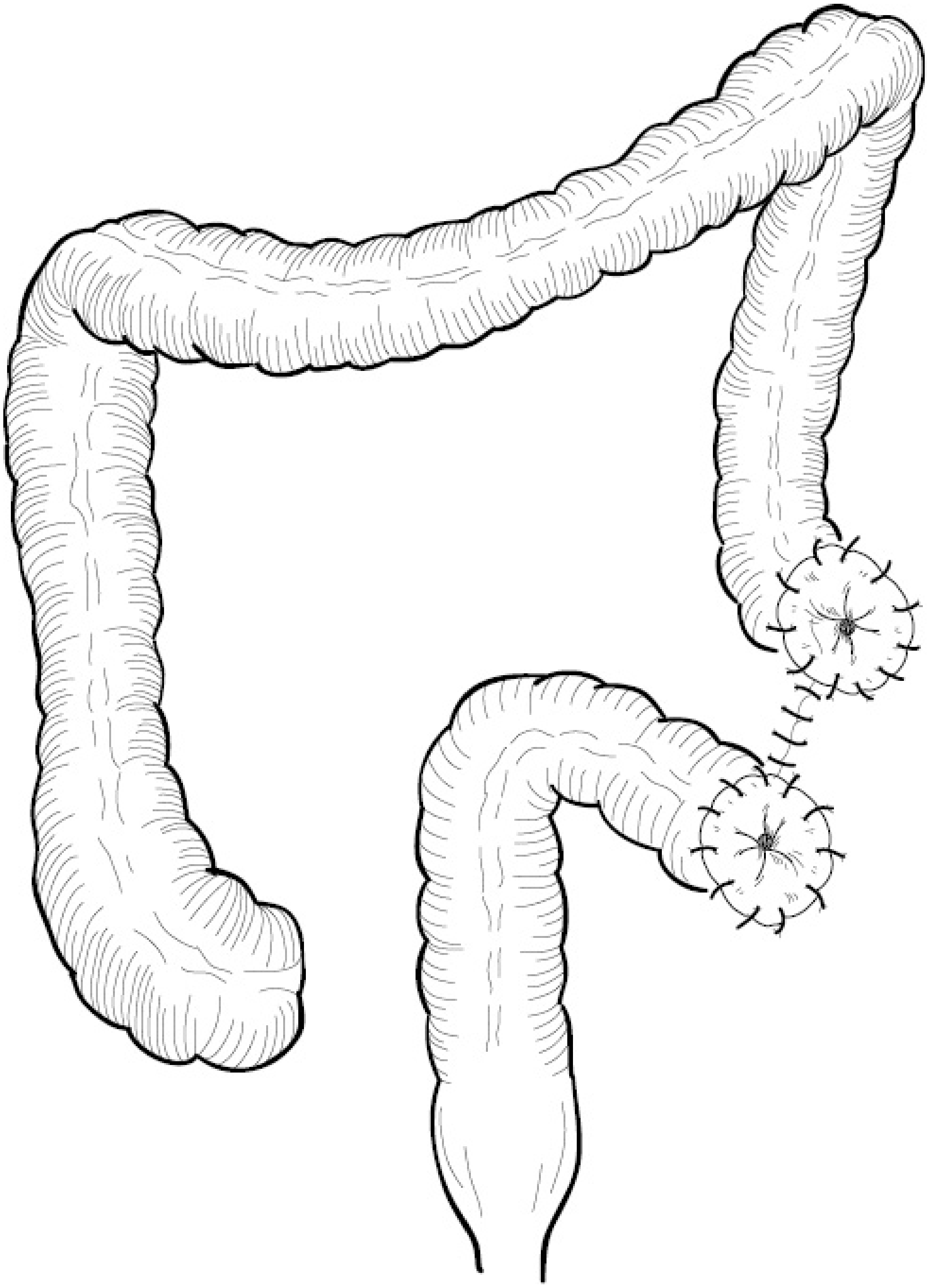
After incising the skin and rectus sheath, the rectus muscle is parted in the line of its fibers; laterally, the abdominal wall muscles are divided if extra space is needed. The peritoneal cavity is entered and the colon identified and exteriorized. This may be difficult when the colon is severely distended and a rectal washout is not possible, particularly as the sigmoid often falls to the right side of the abdomen. If locating the sigmoid colon is difficult, consideration should be given to performing a formal laparotomy.

A transverse colostomy is placed toward the right side of the transverse colon (**Figure 51.2**). If the sigmoid colon is distended, it may lie in the upper abdomen and must be distinguished from the transverse colon by the presence of the fatty tenia coli, which are not found on the transverse colon.



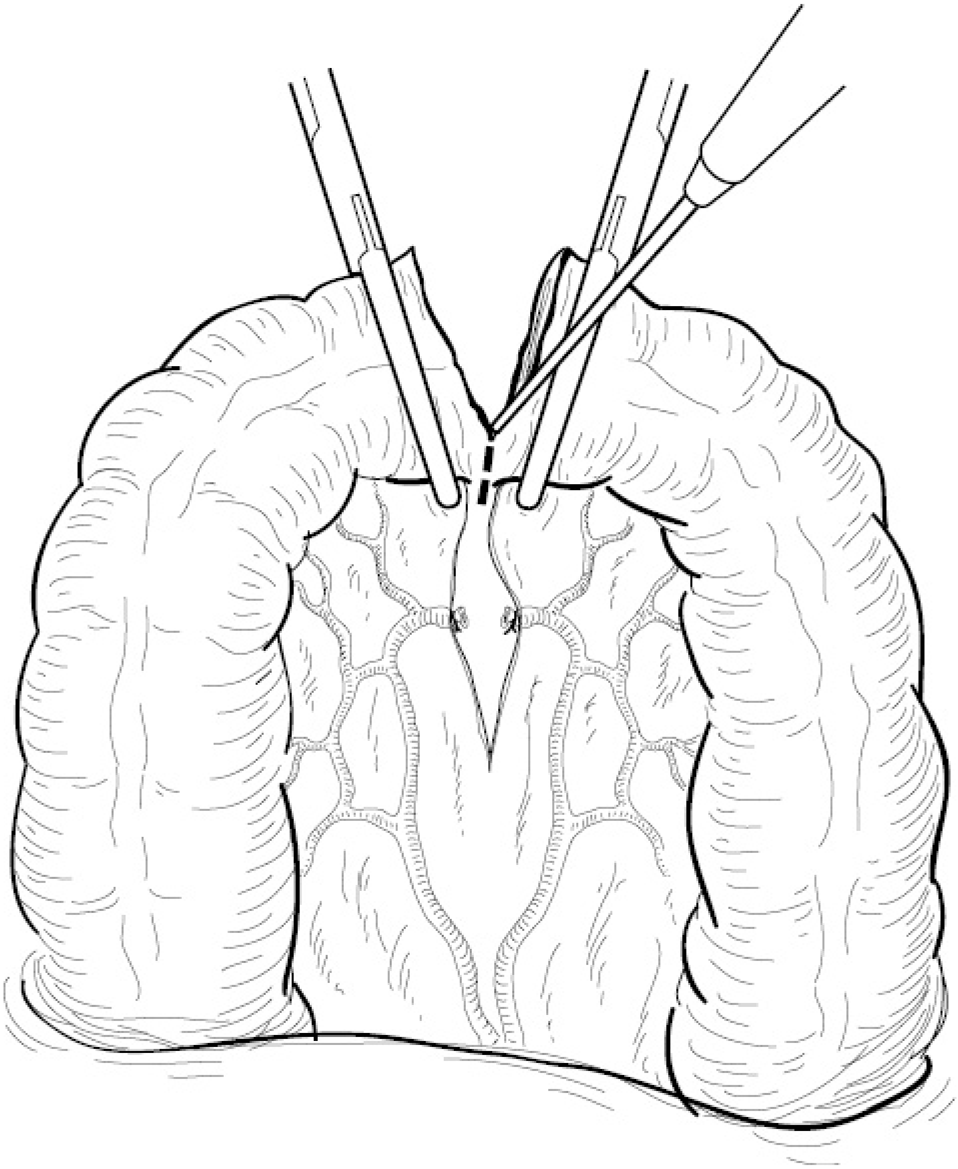
51.2

A sigmoid colostomy is usually placed nearer the proximal sigmoid colon, particularly for neonates with anorectal anomalies so as not to compromise the subsequent pull-through procedure (**Figure 51.3**). For Hirschsprung disease, the stoma is sited proximal to the transition zone as determined by intraoperative seromuscular biopsy. A transverse colostomy is recommended for cloacal anomalies to allow sufficient distal colon for vaginal reconstruction.



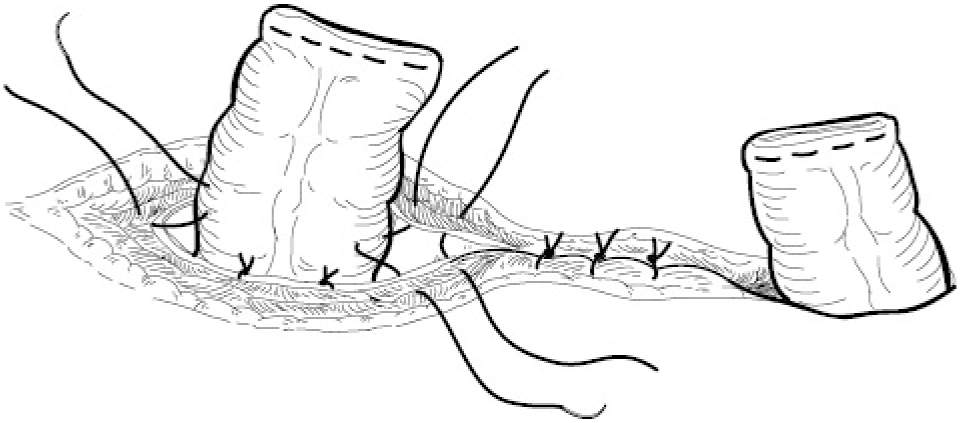
51.3

At the proposed point of transecting the colon, the marginal artery is divided between ligatures, and smaller branches to the colon are coagulated using bipolar diathermy. The colon is then divided using a scalpel or diathermy (**Figure 51.4**). Alternatively, a GIA stapler may be used in older children.



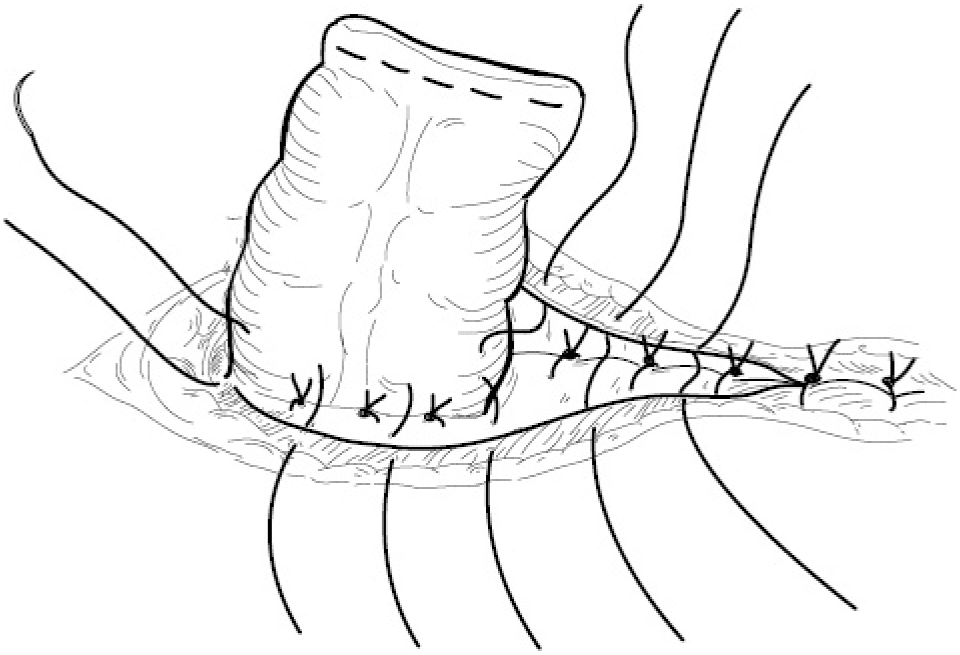
51.4

The proximal and distal limbs of the colon are positioned at opposite corners of the incision, with sufficient distance between them to enable a colostomy bag to enclose the proximal stoma. Each limb is anchored to the abdominal wall with interrupted absorbable sutures in layers (**Figure 51.5**). The first layer is to the colon, peritoneum, and transversalis fascia; these sutures must be sufficiently close together to prevent loops of small intestine from prolapsing between them and should not penetrate the colonic mucosa as this may result in a fecal fistula.



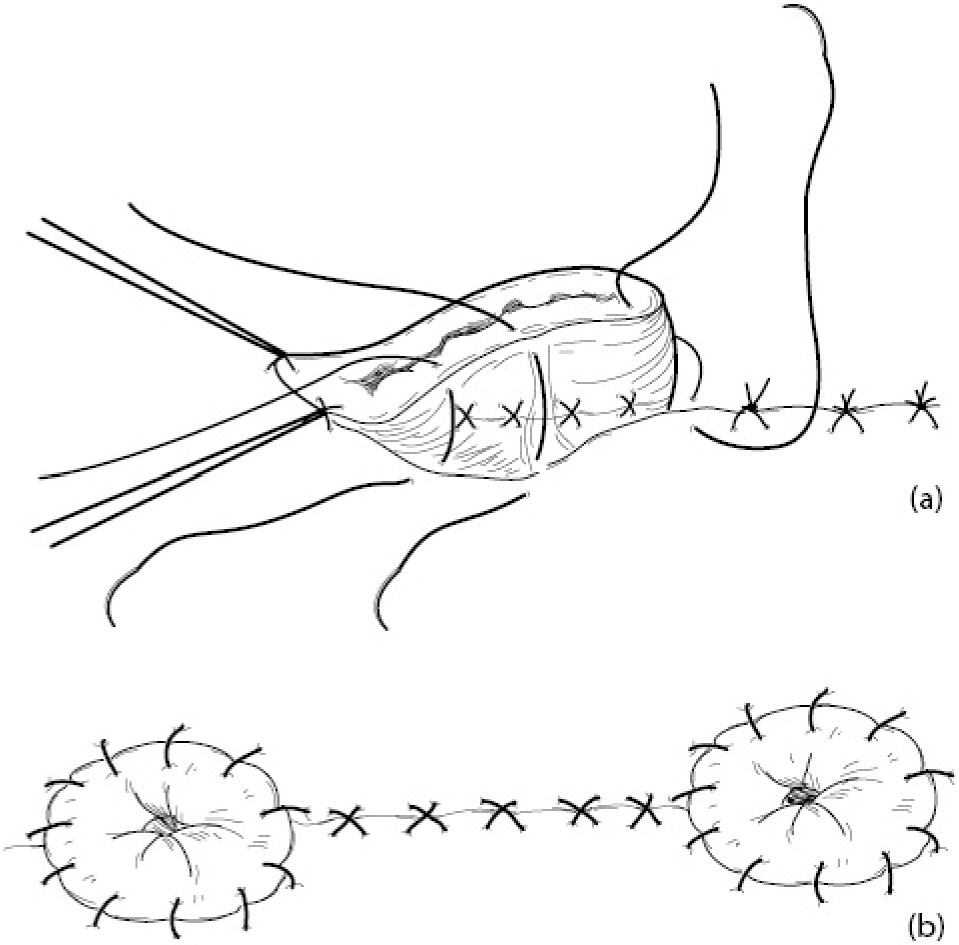
51.5

For the second layer, the colon is sutured to the surrounding external oblique aponeurosis, taking care not to constrict the colon. The abdominal wall layers are approximated between the two limbs of the colostomy (**Figure 51.6**).



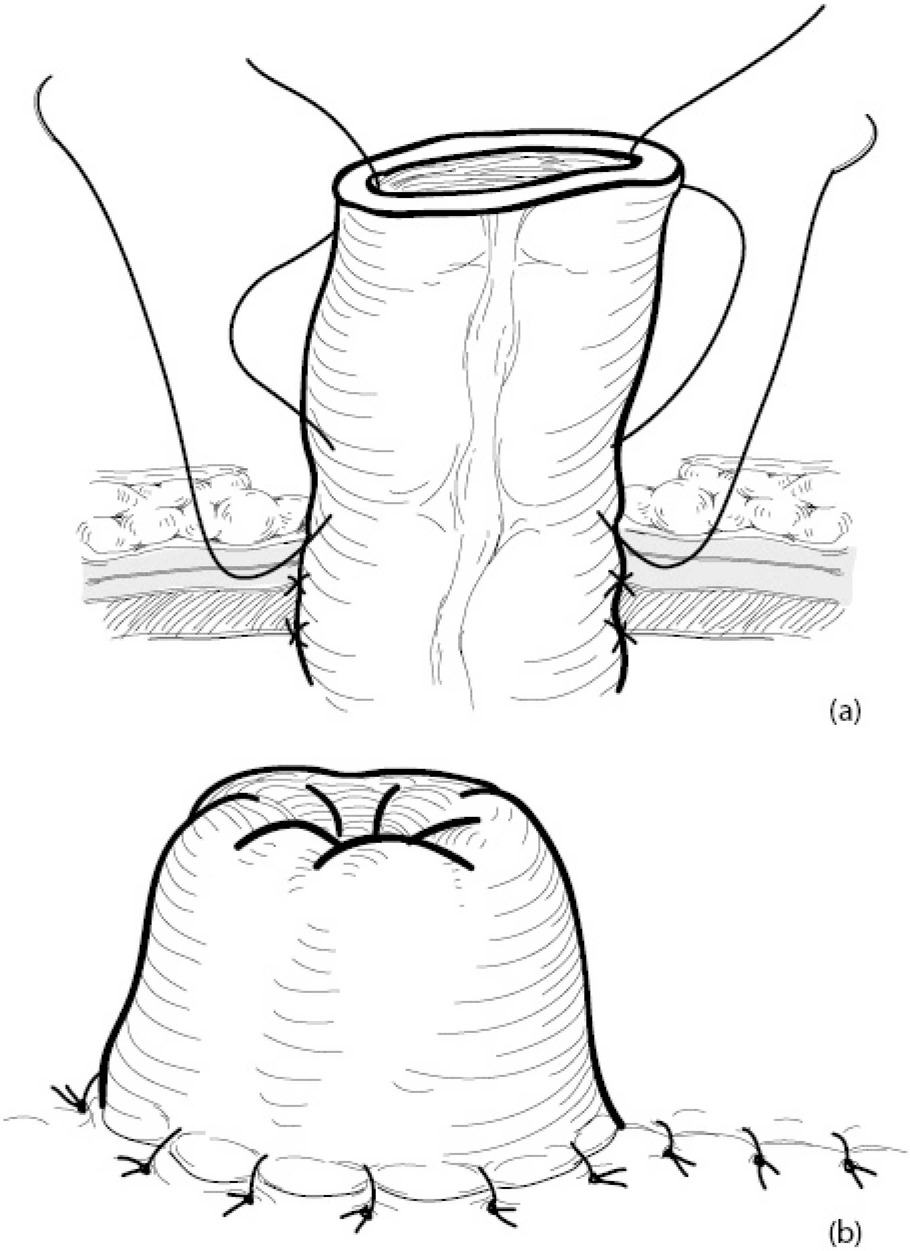
51.6

Each limb of the colostomy is then opened (**Figure 51.7a**). Full thickness, interrupted sutures approximate the cut edge of the colon to the adjacent edges of the skin incision (**Figure 51.7b**).



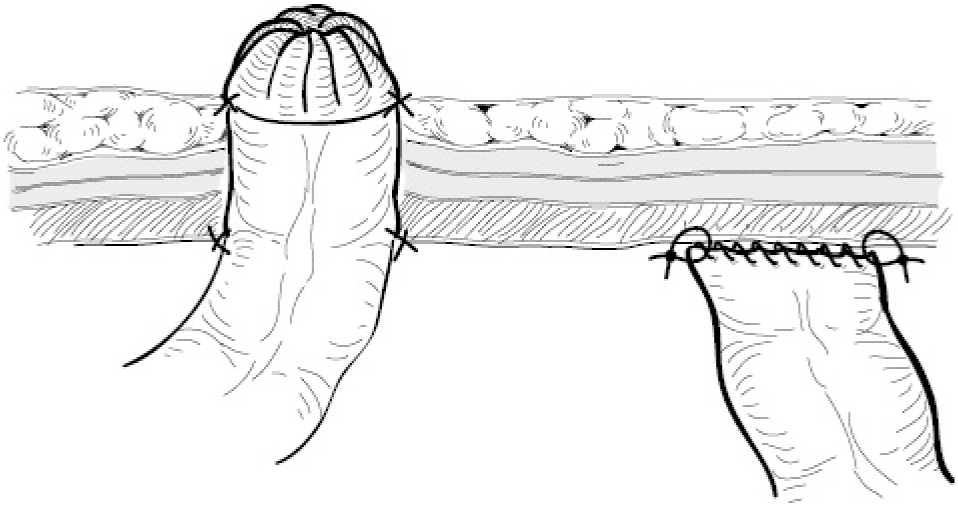
51.7a,b

If required, the proximal stoma may be constructed with a spout that will protrude into the colostomy bag (**Figure 51.8a**). Three-point interrupted sutures are placed through the abdominal wall muscle, the seromuscular wall of the colon, and the cut edge of the colon to evert the end of the colostomy (**Figure 51.8b**).



51.8a,b

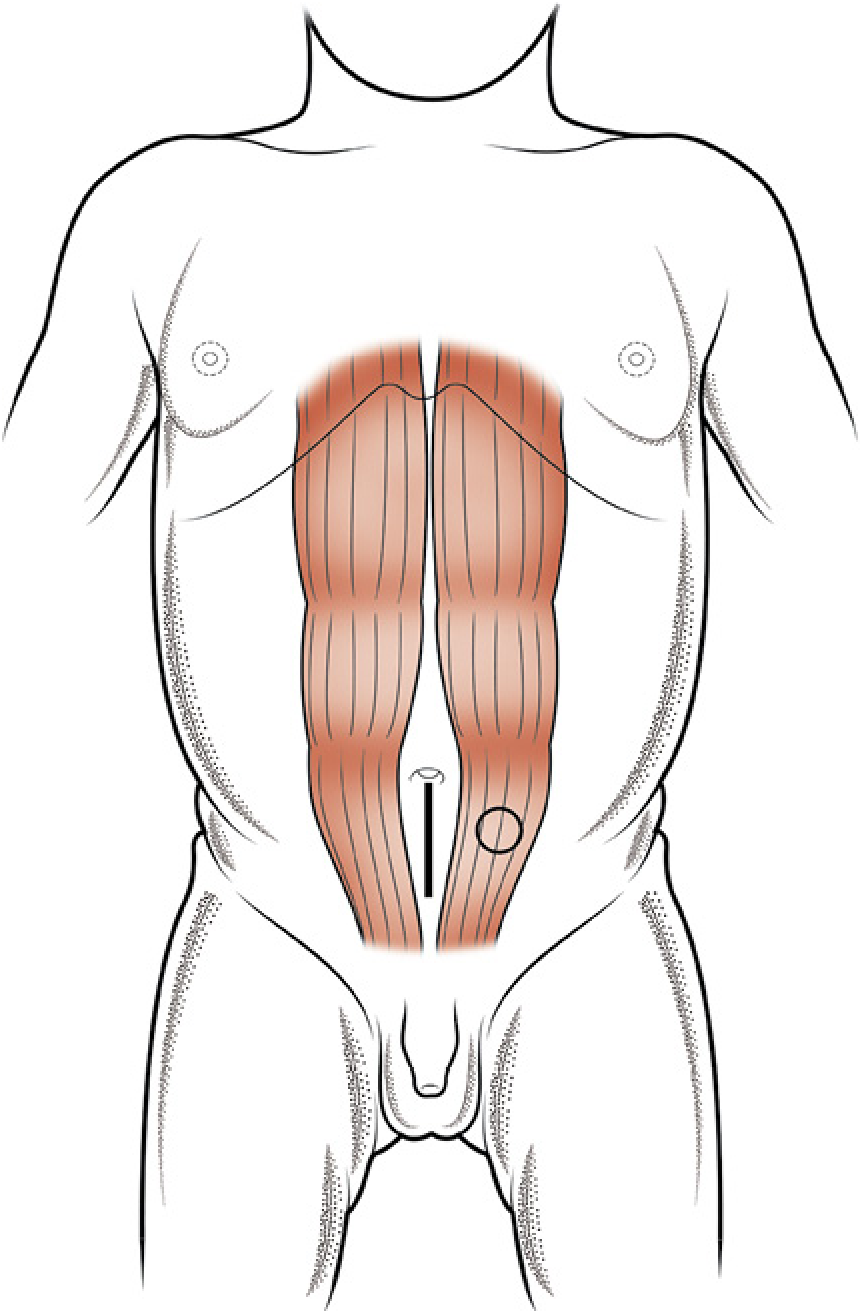
Where there is no obstruction distal to the colostomy, the distal limb may be closed transversely in two layers and placed within the peritoneal cavity (Hartmann procedure). To enable the blind end to be located easily for subsequent reconstruction, it should be anchored to the undersurface of the peritoneum when the wound is closed (**Figure 51.9**).



51.9

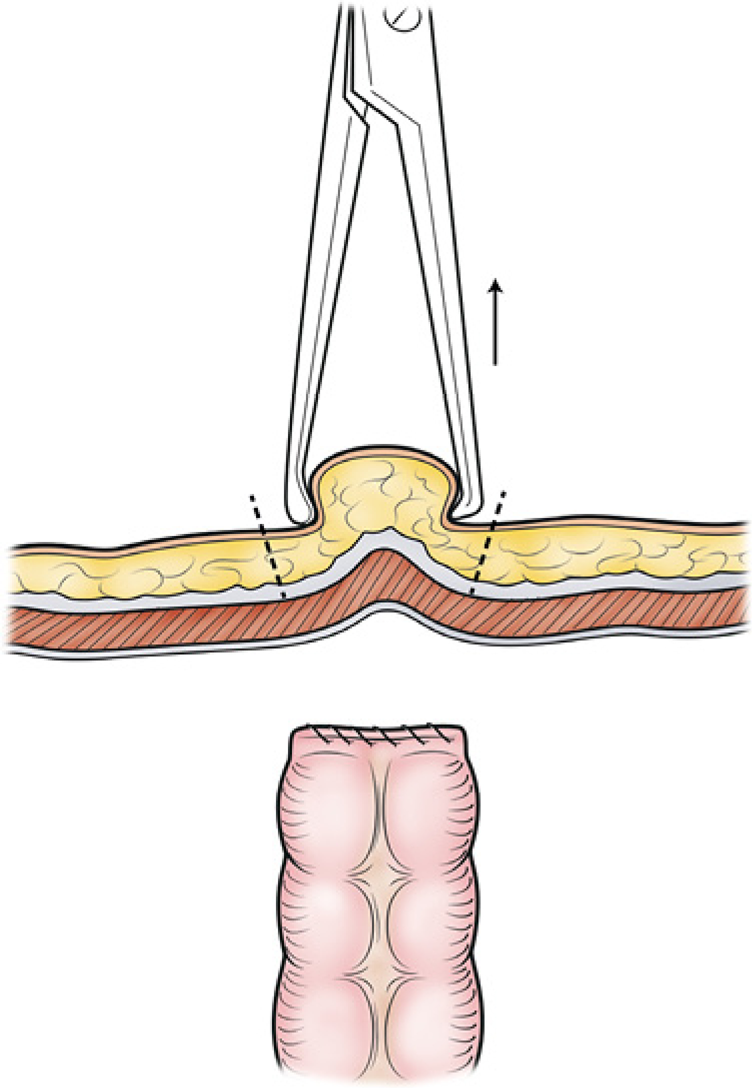
Single-end colostomy

Once the stoma site has been selected based on patient characteristics and underlying pathology, an incision *separate* from the stoma site is made to allow the desired part of the colon to be selected, mobilized, and divided using a stapler device such as an Endo GIA™ (Covidien, Mansfield, MA, USA) (**Figure 51.10**). The distal end of the divided bowel is oversewn with monofilament suture and dropped back into the peritoneal cavity.



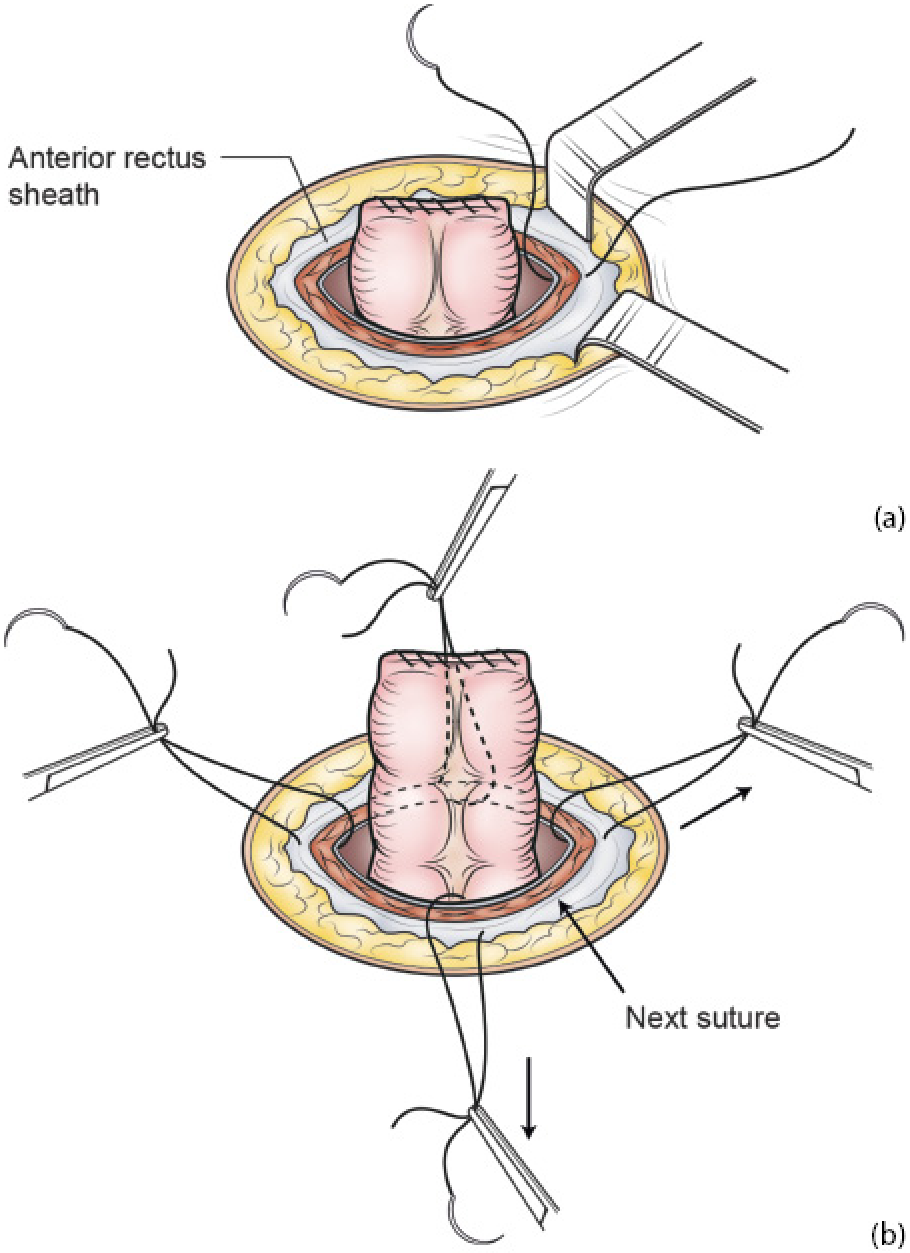
51.10

The center of the marked stoma site (overlying the rectus muscle) is grasped with Allis forceps and a circular incision made to include the skin, fat, and the anterior rectus sheath, and are all excised as a single column (**Figure 51.11**). The rectus muscle is split in the line of its fibers using two Langenbeck retractors to reach the posterior rectus sheath, which is incised transversely. The size of the aperture should match the caliber of the bowel brought out at that point.



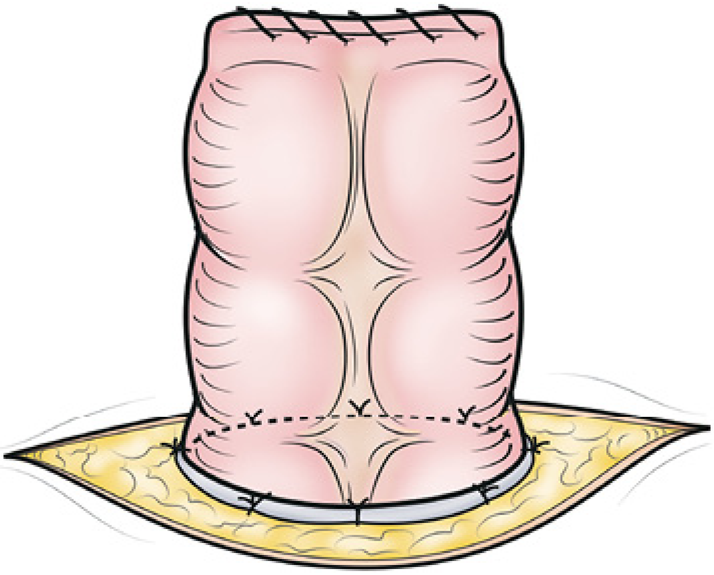
51.11

The closed proximal limb of the bowel is brought out into the circular incision with no contamination, with the mesenteric orientation affirmed through the laparotomy wound (**Figure 51.12a**). To start, four sutures are taken on the anterior and posterior rectus sheaths at four corners using 3/0 or 4/0 Vicryl™ (**Figure 51.12b**). These sutures are put on clips and left untied and needle uncut. The retraction of any two adjacent sutures facilitate another suture placement in between.


Anterior rectus sheath
(a)
Next suture
(b)

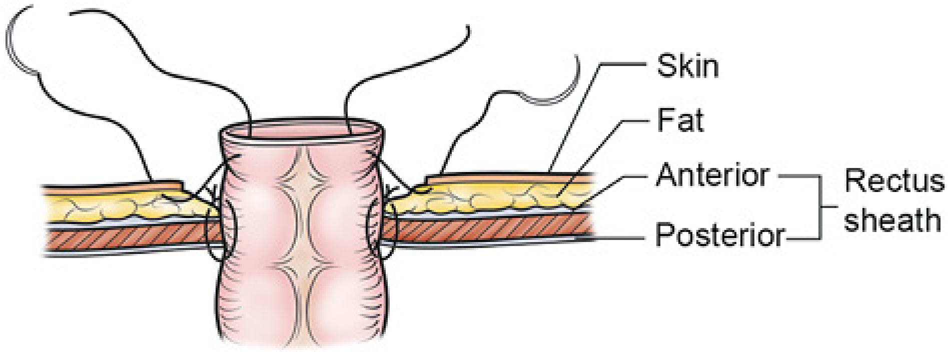
51.12a,b

About eight sets of circumferential rectus sheath sutures may be needed at the same level of the colon, by taking a partial thickness bite on the bowel, and tied. The laparotomy wound is then closed in layers and sealed with tissue glue (**Figure 51.13**).



51.13

The staple line on the bowel is excised, and the colostomy is matured with twelve 3/0 or 4/0 Vicryl sutures. A "pseudospout" is created by taking a relatively larger full thickness bite into the bowel lumen in the first four sutures (**Figure 51.14**).

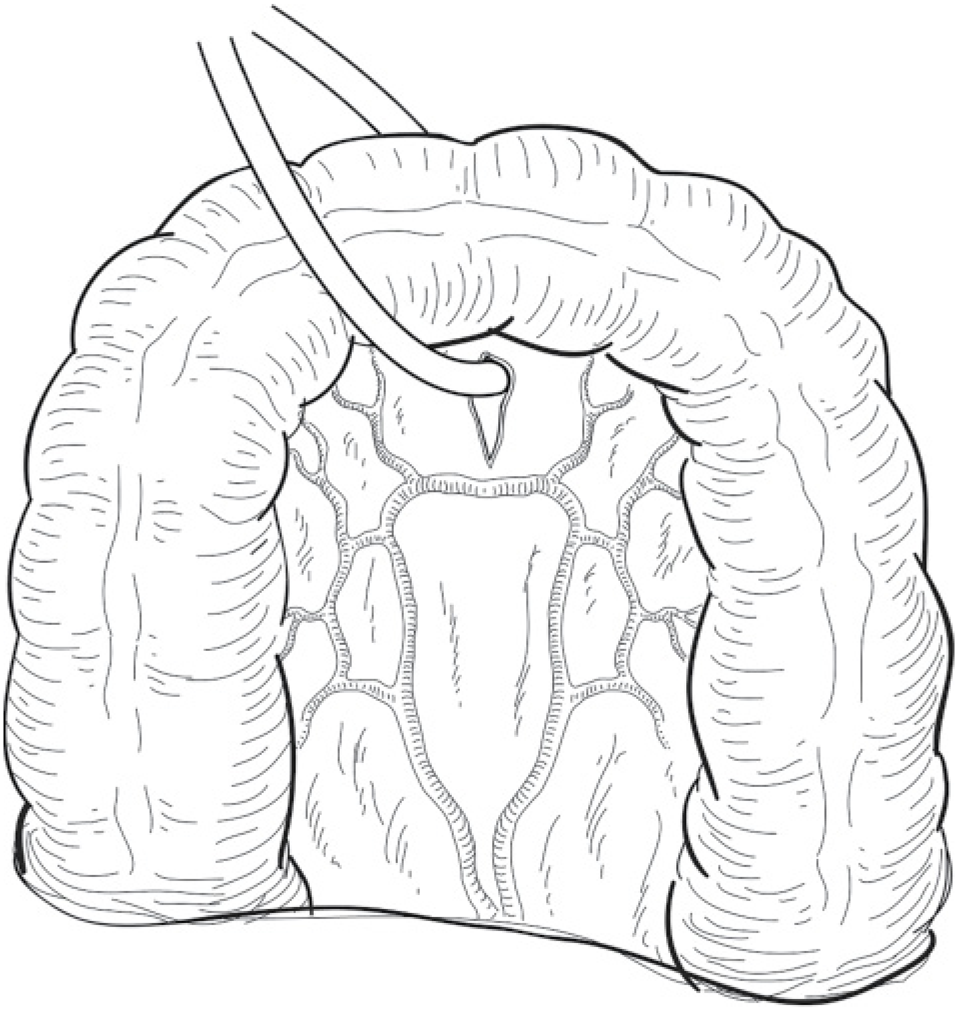

Skin
Fat
Anterior
Posterior
Rectus sheath

51.14

Loop colostomy

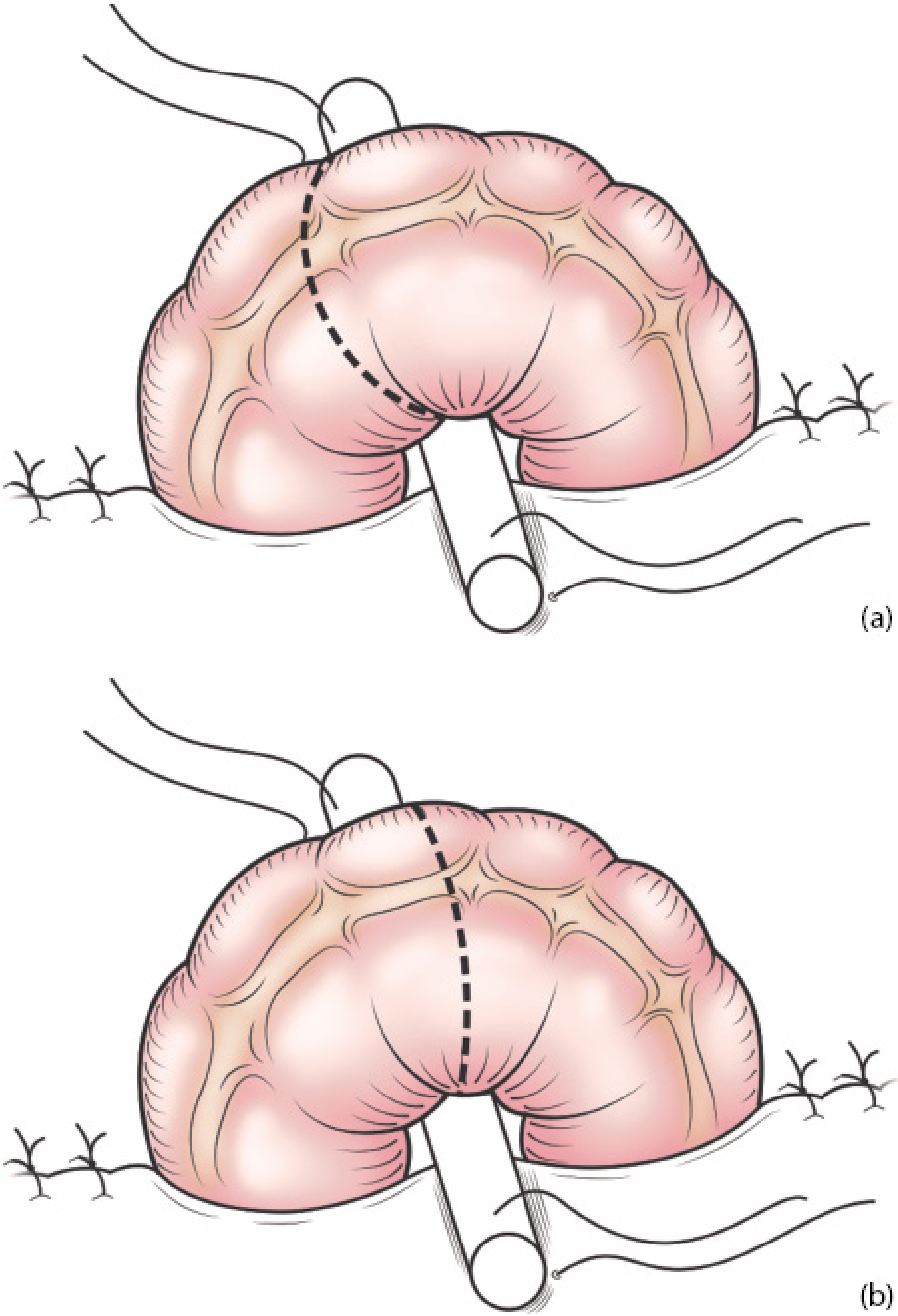
A muscle-splitting incision is used as for a divided colostomy, but the shape of the skin incision will depend on whether the exteriorized colon is to be supported by a catheter tube, in which case a standard straight skin incision is used (**Figure 51.1**), or with a skin flap, for which an inverted V-incision is made (**Figure 51.19**).

The colonic loop is exteriorized and elevated using a soft catheter passed through a window in the mesentery at the point of the proposed stoma (**Figure 51.15**). The mesenteric artery is not divided. The two limbs of the colon are secured to the abdominal wall in layers using absorbable sutures, as described for a divided colostomy. The space between the two limbs is closed with interrupted sutures to prevent the small intestine prolapsing between them, taking care to avoid damaging the mesenteric blood vessels.



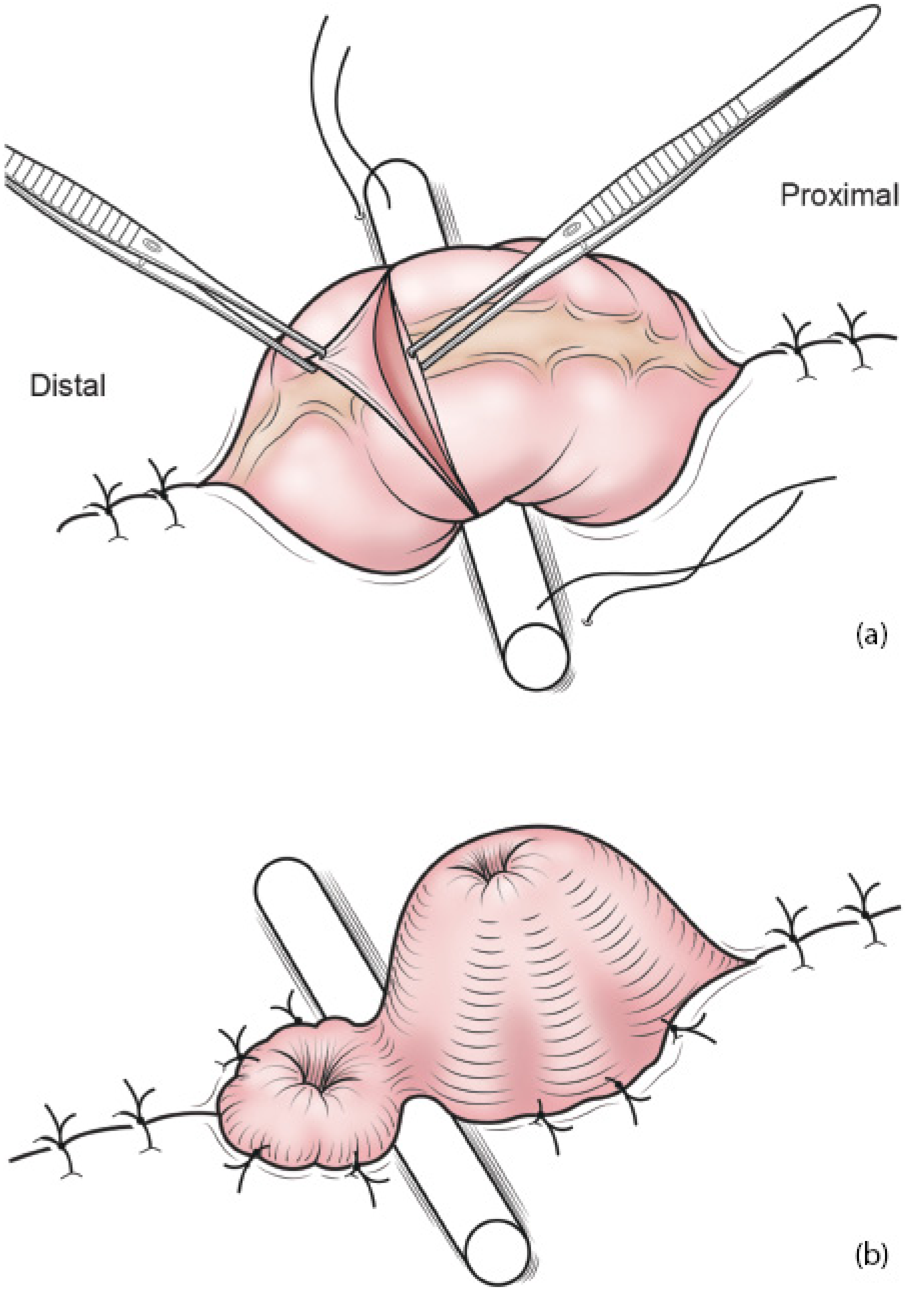
51.15

The catheter is shortened and anchored to the skin on either side of the loop of colon using non-absorbable sutures. The catheter must be short enough to be enclosed by the colostomy bag. The stoma is opened through a transverse incision (**Figure 51.16a**) at the apex of the colonic loop; alternatively, if the desired effect is to make the proximal end wider than the distal end (e.g. in anorectal malformation), an incision deliberately curved distally (**Figure 51.16b**) can be employed instead. To achieve this safely, a short incision is first made so that the edges of the colon can be lifted to avoid damaging its closely opposed opposite wall, particularly if diathermy is used to incise the colon.



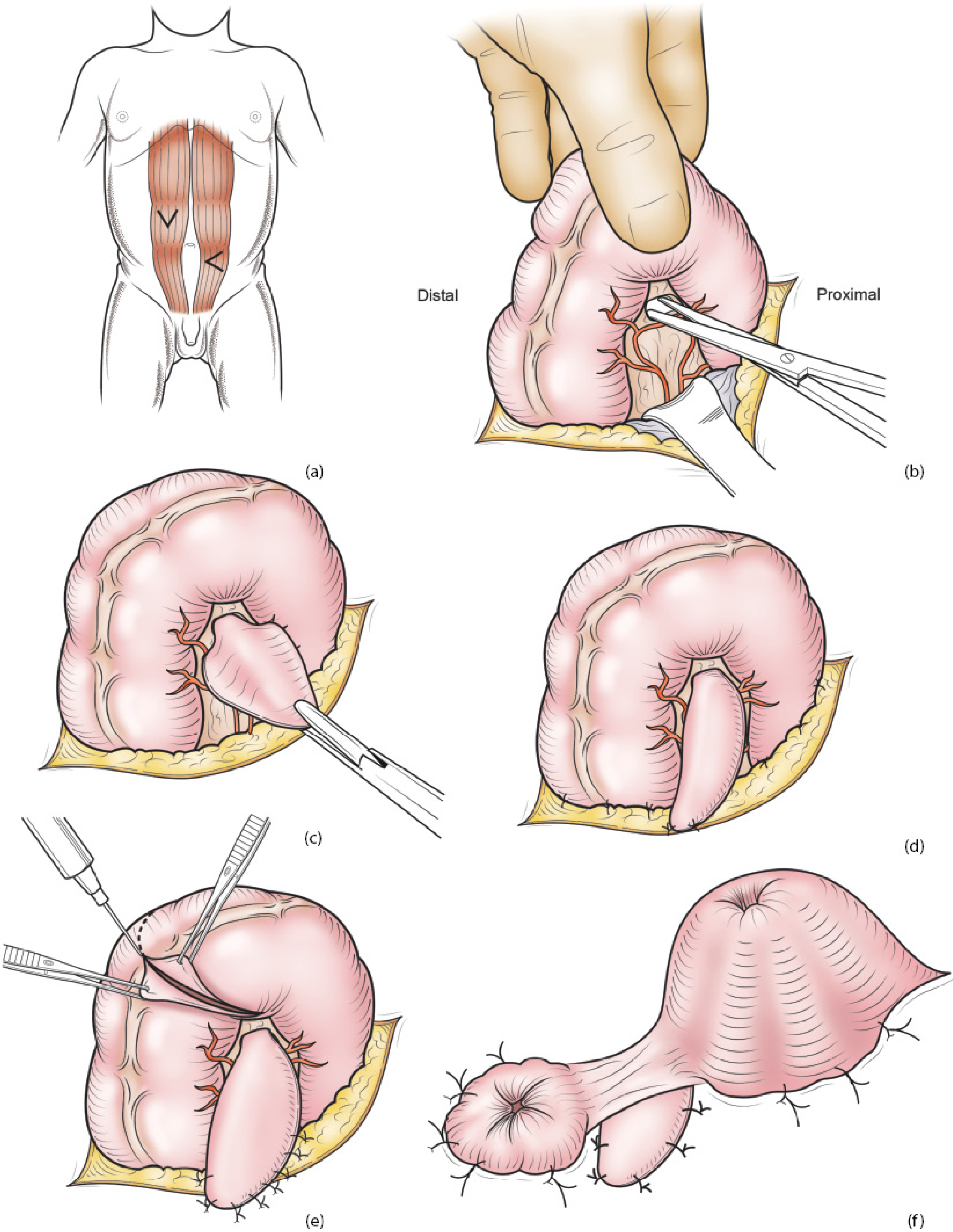
51.16a,b

The edges of the stoma are everted and sutured to the skin incision (**Figure 51.17a and b**). A Hegar dilator is used to confirm that the stoma has not been constricted.



51.17a,b

If a skin flap is used to support the colostomy, the flap is drawn through the window in the mesentery and sutured as illustrated in **Figure 51.18a–f**. If a larger incision is required to examine or orientate the bowel, extension from one end or apex of the V can be made, and then closed just short of the skin flap.


Distal
Proximal
(a)
(b)
(c)
(d)
(e)
(f)

51.18a–f

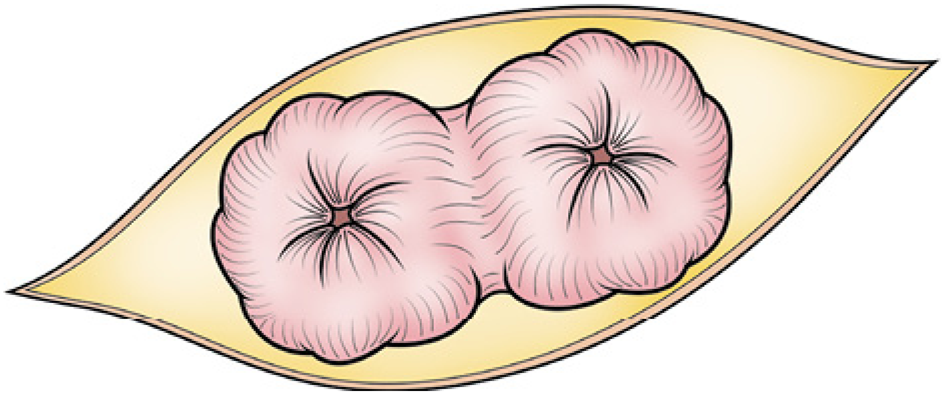
Intraoperative colostomy

If a colostomy is required following intestinal resection, it should ideally be brought out through a separate incision. An exception is when the remaining intestine is fragile and compromised (as in neonatal necrotizing enterocolitis) and exteriorizing the colonic loops through separate incisions would be traumatic and risk further loss of valuable intestinal length. In this situation, the colonic loops may be brought out at either end of the main abdominal incision and carefully anchored to the abdominal wall. It is not always necessary to evert and "mature" the stoma as this will occur spontaneously.

Colostomy closure

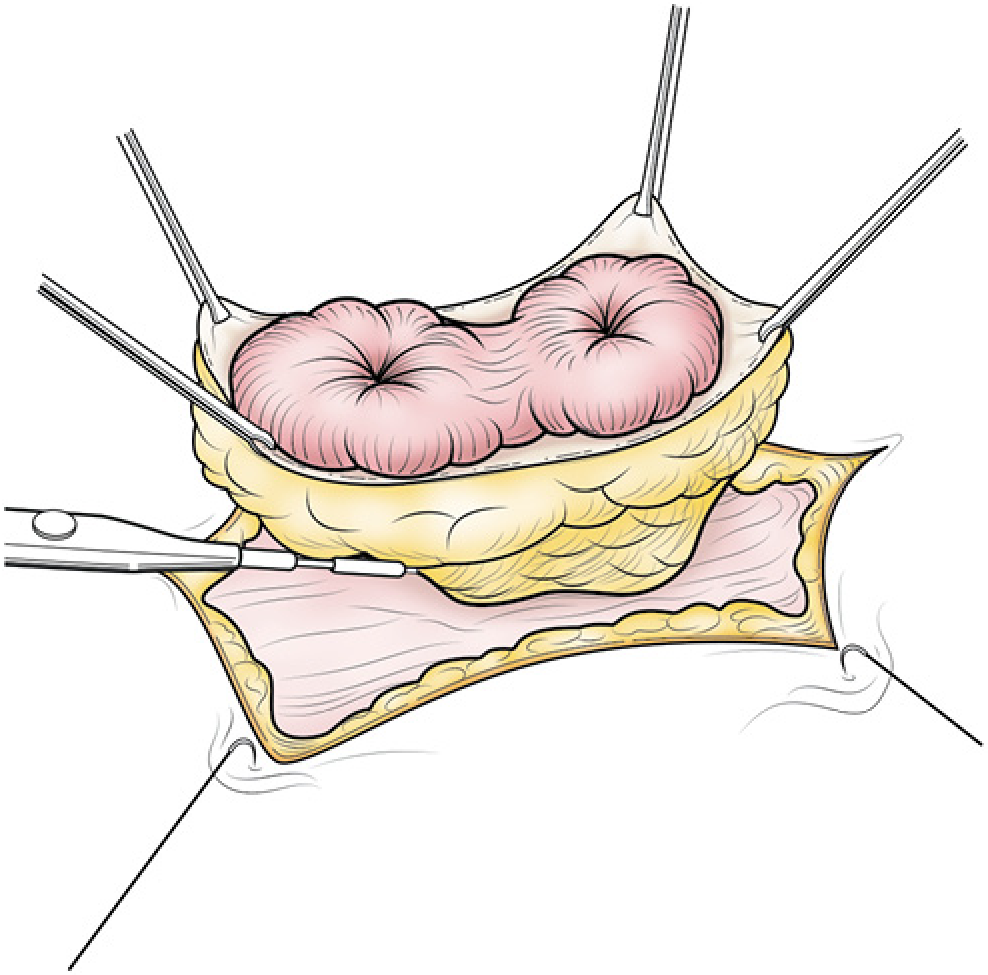
Prior to closing the colostomy, the integrity and patency of the bowel distal to the stoma must be confirmed by preoperative contrast radiology and, if appropriate, by digital rectal examination at the time of operation. Bowel preparation to empty the colon and rectal irrigation to evacuate residual stool may be necessary. Clear enteral fluids for 24 hours preoperatively can be used to reduce fecal residue.

The incision should be elliptical and includes the scar tissue (**Figure 51.19**). The skin should be incised in stages as there may be brisk arterial oozing from the margins of the incision. Monopolar diathermy can be employed for the skin incision to minimize blood loss if the skin is particularly inflamed.



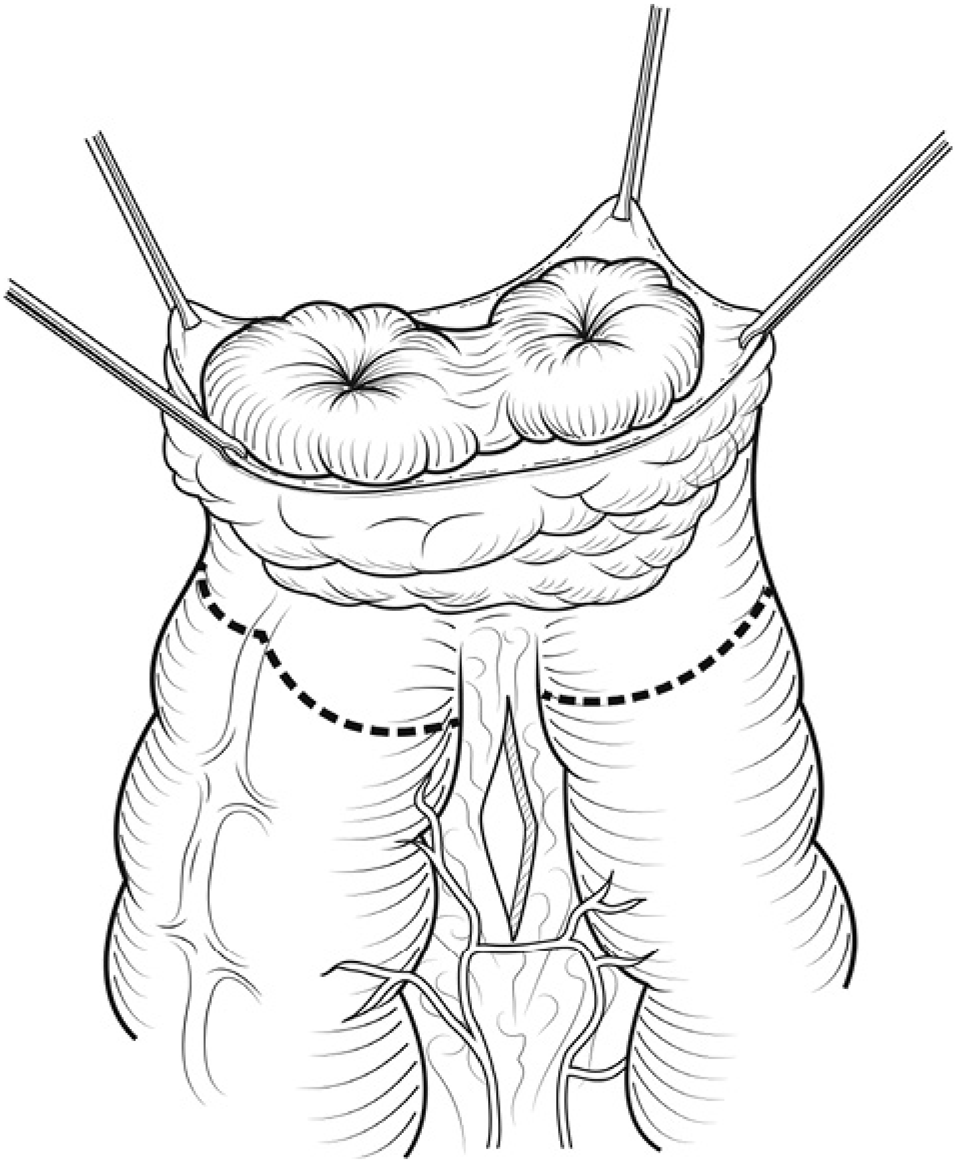
51.19

The incision is deepened to expose the external oblique aponeurosis and the margins of the opening in the abdominal wall are identified. With careful dissection using a combination of sharp dissection and monopolar diathermy, a plane is developed between the colonic loop and the abdominal wall to enter the peritoneal cavity, dividing the anchoring sutures and inevitable scarring without breaching the colonic wall or damaging the adjacent adherent loops of small intestine (**Figure 51.20**). This may be a difficult process and patience is essential. With a divided colostomy, the incision between the stomas is opened completely.



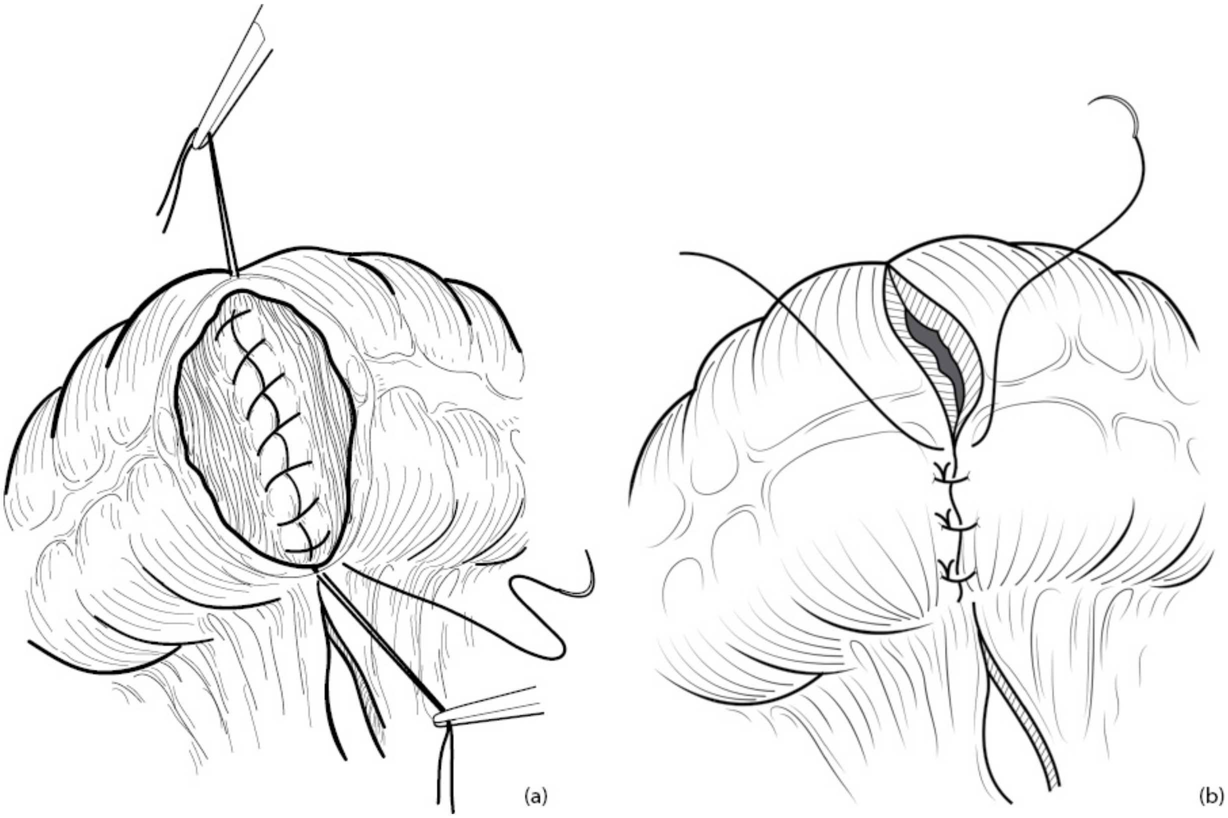
51.20

Once the two limbs of the colostomy have been mobilized sufficiently to allow them to be exteriorized, the stomas are resected back to healthy bowel and approximated by end-to-end anastomosis (**Figure 51.21**). If possible, care should be taken to preserve the marginal vessels.



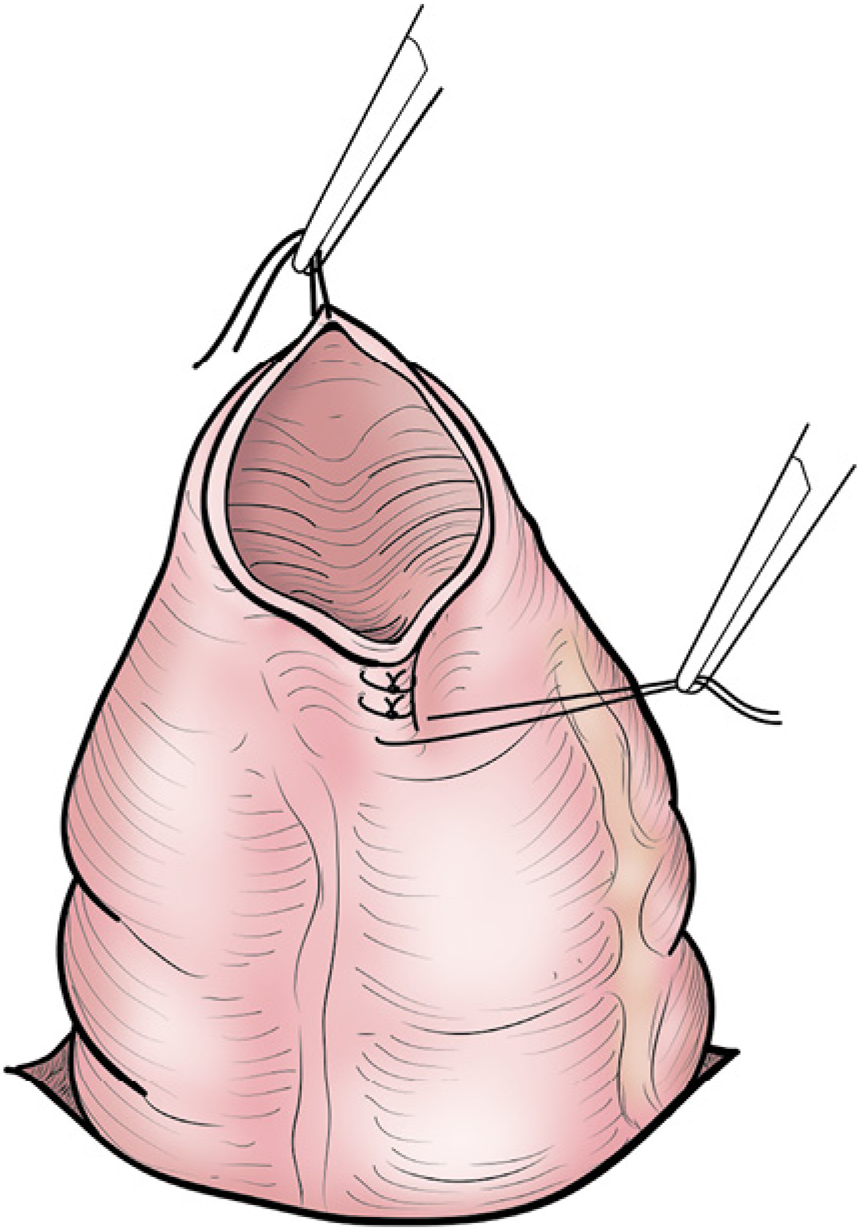
51.21

A single layer of interrupted extramucosal sutures is suitable for all ages (**Figure 51.22a**). Once the anastomosis is completed, the mesenteric defect is closed with interrupted sutures if wide (**Figure 51.22b**).



51.22a,b

In the older child, a loop colostomy may be closed without resecting the stoma (**Figure 51.23**). After mobilizing the colostomy, the edges are trimmed and the colon is closed transversely. In the infant, resection and end-to-end anastomosis are recommended.



51.23

POSTOPERATIVE CARE

Postoperatively, IV fluids are continued until an adequate oral fluid intake is tolerated. Nasogastric tubes are not always required postoperatively, especially in infants. Enteral feeds are commenced usually within 24 hours and increased cautiously until flatus or stools are passed. Systemic analgesia is maintained as required.

COMPLICATIONS

Immediate complications of colostomy formation include uncontrolled intra-abdominal spillage, reverse orientation of the colostomy with the mucous fistula, and colostomy formed under tension causing retraction, all potentially preventable with meticulous attention to detail. Following colostomy formation, colostomy-related complications have been reported in up to 80% of patients. These include peristomal skin excoriation, prolapse, stomal obstruction/stenosis, parastomal herniation, and stomal bleeding.

Peristomal excoriation is very common and requires meticulous attention to skin care using specific protective preparations under the supervision of a stoma therapist. Prolapse from either loop may be troublesome, requiring repeated manual reduction and, occasionally, stomal revision. Stomal stenosis resulting in intestinal obstruction may require dilatation or revision. Parastomal hernias occur as a result of small intestine herniating between the sutures anchoring the colostomy to the abdominal wall. Diversion proctitis, characterized by a mucopurulent rectal discharge, bleeding, and tenesmus, is a late effect of colonic diversion with characteristic endoscopic and histological features.

Early complications following colostomy closure include wound infection, the risk of which is reduced by prophylactic antibiotics, and anastomotic leak. The latter is uncommon provided the bowel is healthy and well-vascularized, there is no tension on the anastomosis, and no distal bowel obstruction. Meticulous operative technique is required to ensure healthy bowel wall edges at the time of anastomosis. Late stenosis at the suture line is also uncommon if the colon is healthy. Adhesive small bowel obstruction has been reported in about 5% of children following colostomy closure, and a mortality of <1%.

VIDEO

OPS_8e_video_51.1 Colostomy construction

FURTHER READING

Antoniou SA, Agresta F, Garcia Alamino JM, et al. European Hernia Society guidelines on prevention and treatment of parastomal hernias. *Hernia* 2018; 22(1): 183–98.

Bischoff A, Levitt M, Lawal TA, Pena A. Colostomy closure: How to avoid complications. *Pediatr Surg Int* 2010; 26: 1087–92.

Cigdem MK, Onen A, Duran H, et al. The mechanical complications of colostomy in infants and children: An analysis of 473 cases in a single center. *Pediatr Surg Int* 2006; 22: 671–6.

Gine C, Santiago S, Lara A, et al. Two-port laparoscopic descending colostomy with separated stomas for anorectal malformations in newborns. *Eur J Pediatr Surg* 2016; 26: 462–4.

Irtan S, Bellaïche M, Brasher C, et al. Stomal prolapse in children with chronic intestinal pseudoobstruction: A frequent complication? *J Pediatr Surg* 2010; 45: 2234–7.

Kiely EM, Ajayi NA, Wheeler RA, Malone M. Diversion procto-colitis: Response to treatment with short-chain fatty acids. *J Pediatr Surg* 2001; 36: 1514–17.

Pena A, Migotto-Krieger M, Levitt MA. Colostomy in anorectal malformations: A procedure with serious but preventable complications. *J Pediatr Surg* 2006; 41: 748–56.

van den Hondel D, Sloots C, Meeussen C, Wijnen R. To split or not to split: Colostomy complications for anorectal malformations or Hirschsprung disease: A single center experience and a systematic review of the literature. *Eur J Pediatr Surg* 2014; 24: 61–9.

Youssef F, Arbash G, Puligandla PS, Baird RJ. Loop versus divided colostomy for the management of anorectal malformations: A systematic review and meta-analysis. *J Pediatr Surg* 2017; 52: 783–90.

52

Surgery of short bowel syndrome: Longitudinal intestinal lengthening and tailoring and serial transverse enteroplasty

RICCARDO COLETTA, SAM M. HAN, CHARLES R. HONG, ANTONINO MORABITO, AND TOM JAKSIC

INTRODUCTION

Intestinal failure in infants and children primarily results from insufficient intestinal mass, i.e. a "short bowel" which is insufficient to maintain adequate nutrition to support normal physiology and growth. Consequent long-term dependency upon parenteral nutrition (PN) may result in significant morbidity and mortality from intestinal failure-associated liver disease (IFALD), catheter-related bloodstream infections (CRBSIs), and metabolic and renal disease.

Early adoption of synergistic medical and surgical strategies to augment the natural physiological process of adaption and maximize the potential for enteral autonomy is critical to long-term survival in this population.

The surgery of short bowel syndrome (SBS) has been designed to maximize the potential of the existing bowel and overcome the problems of proximal intestinal dilatation, impaired antegrade motility, and bacterial sumping and overgrowth. Modern surgical treatment can be said to have begun in 1980 when Adrian Bianchi designed the first effective method of bowel lengthening, now known as longitudinal intestinal lengthening and tailoring (LILT). This improved the chances of enteral autonomy and reduced the requirement for PN. An alternative procedure, serial transverse enteroplasty (STEP), was first described in a porcine model in 2003 and has also gained wide acceptance by pediatric surgeons both for the initial reconstruction of dilated bowel and for repeat surgery for redilation. The first human STEP was performed on a 2-year-old with SBS from gastroschisis and midgut volvulus who had previously undergone LILT.

LONGITUDINAL INTESTINAL LENGTHENING AND TAILORING (LILT)

PRINCIPLES AND JUSTIFICATION

The medical management of SBS has changed considerably in the last three decades most notably in the adoption of more sophisticated PN regimens with lipid restriction and/or omega-3 emulsions have considerably diminished the risks of IFALD particularly. This has been allied to intestinal rehabilitation programs where a multidisciplinary expertise offers a tailored treatment for each patient. Nonetheless, even with appropriate medical management, surgical intervention is still considered an important tool to improve survival and reduce morbidity.

The principle behind LILT is the creation of two hemiloops from a single dilated loop of small bowel that are then anastomosed sequentially. Bowel length therefore increases (80–100%) at the expense of bowel dilatation. Clearly, the absorptive surface area has not changed, but nutrient contact with mucosa increases to improve functionality.

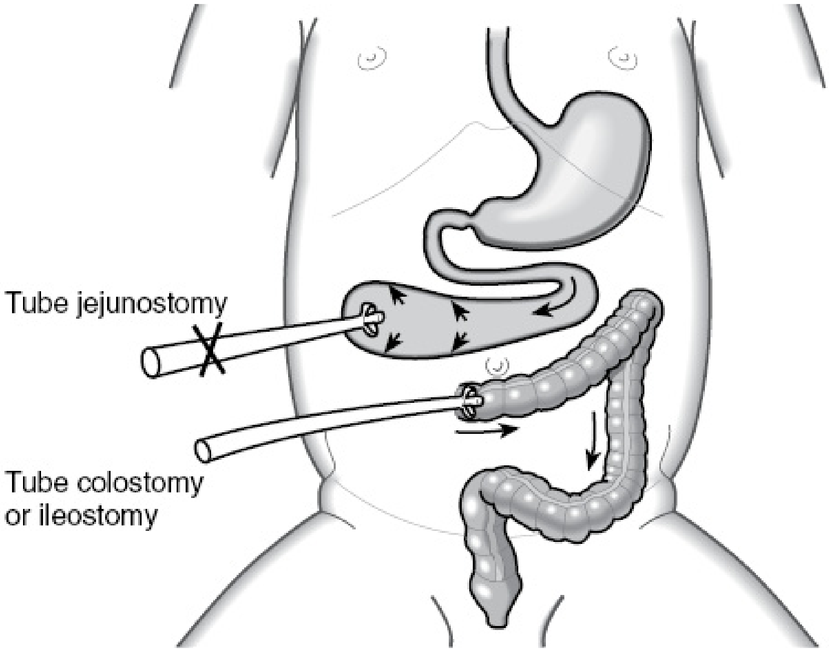
LILT is indicated in patients with <40 cm of remnant bowel, with a dilated segment more than 5–6 cm and with a non-sclerotic mesentery. It has been used also in children with a more fibrotic mesentery typically as a consequence of necrotizing enterocolitis (NEC).

In Manchester, LILT is considered in PN-dependent children who are not achieving at least 50% of their calorie requirement enterally after 6 months of adequate medical treatment.

PREOPERATIVE INVESTIGATIONS

Careful evaluation of remnant intestinal dilatation is the key to determine whether a LILT procedure is possible.

In selected neonates with extensive midgut loss and residual jejunal atresia, we have adopted a strategy to maximize proximal expansion using *controlled jejunal* *expansion*. At the initial surgery shortly after birth we place a Foley (6 Fr or 8 Fr) or a Malecot (16 Fr) catheter into the proximal bowel and bring it out through the abdominal wall. A second catheter is also placed in the distal bowel (usually colon) to allow for recycling of jejunostomy losses, to maximize absorption, and to stimulate colonic adaptation (**Figure 52.1**).



52.1

This system allows free drainage of the proximal bowel and facilitates early enteral feeding, thus stimulating the child to become "food-wise" and inducing small-bowel mucosal adaptation. We strongly recommend oral rather than nasogastric or gastrostomy feeding to inhibit food aversion, common in children with SBS. Intermittent occlusion of the proximal tube stoma preserves and stimulates further dilatation.

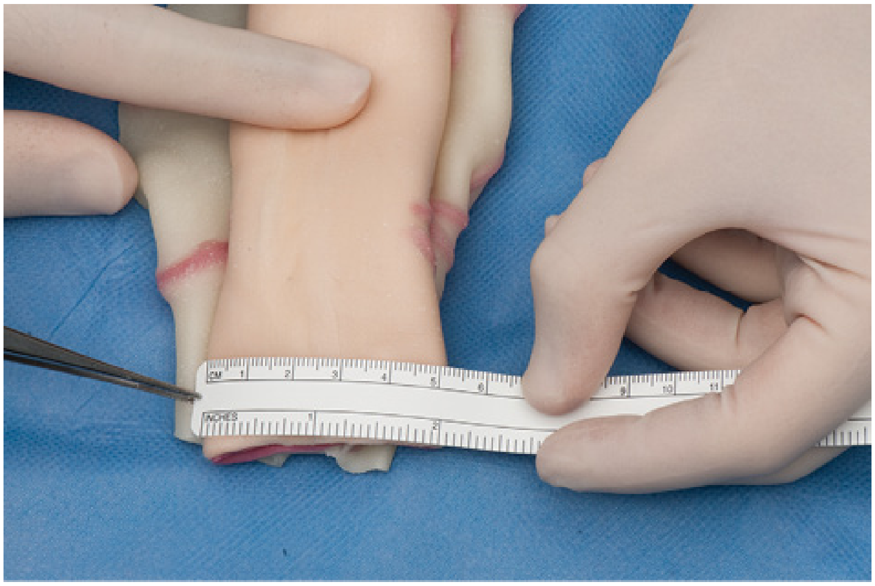
In SBS patients without stoma, a water-soluble contrast follow-through should be completed to understand dilatation and length of residual bowel. Liver and kidney ultrasonography should be used for early detection of IFALD and nephrocalcinosis respectively. Doppler flow studies of the neck and groin establish a "road-map" of venous patency and pre-existing thrombosis resulting from previous central venous catheters.

Current hematology, coagulation, liver biochemistry, and nutritional indices should be available in all SBS patients.

OPERATION

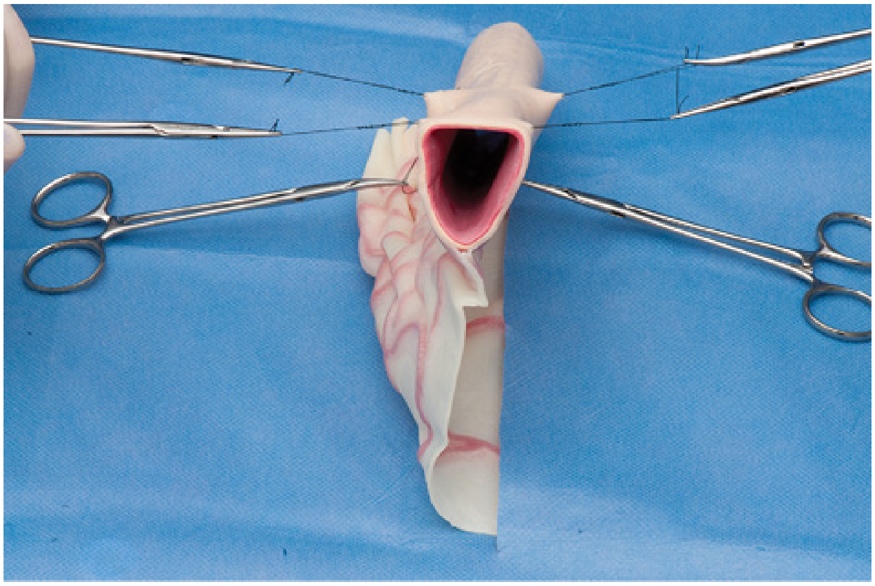
The abdomen is opened through an already existing scar or through a transverse subumbilical incision. Where stomas are present, they are taken down at this stage. The small and large bowel is freed with meticulous attention to avoid damage to the bowel and to the vascular supply. Previous anastomosis between the dilated proximal bowel (to be lengthened) and the distal bowel (often the colon) are taken down.

Measurement of the remnant bowel length is performed employing a soft tie that runs over the antimesenteric border. Dilatation of the bowel to be lengthened is measured by flattening the segment on a damp swab and use of a ruler (**Figure 52.2**).



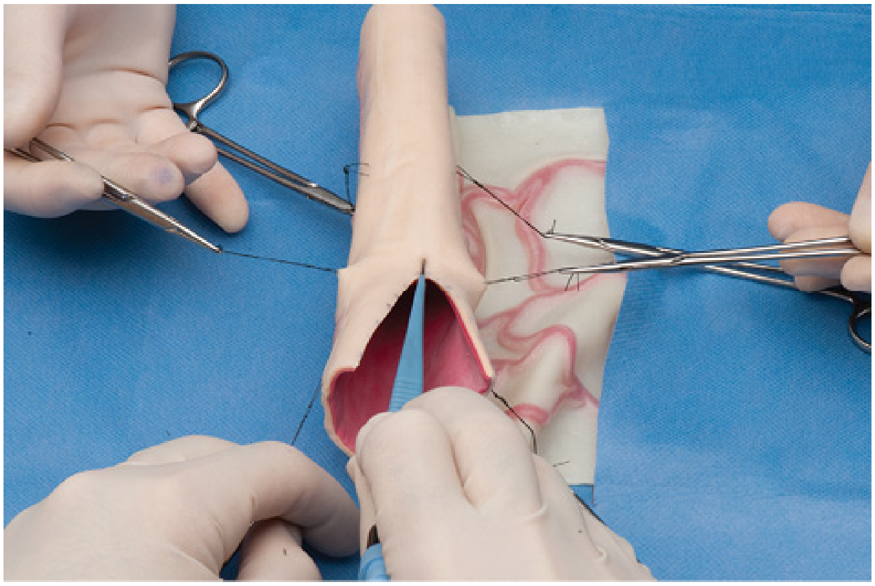
52.2

Stay sutures (silk 4/0) are placed at the beginning and at the end of the selected segment to be lengthened. Traction sutures (Maxon□ 4/0) are placed along the antimesenteric border to the right and the left of the midline at about 5 cm intervals. A gentle upward and outward lift against the base of the mesentery is done holding the traction suture (**Figure 52.3**). This maneuver allows the surgeon to focus the attention on the entire midline of the bowel.



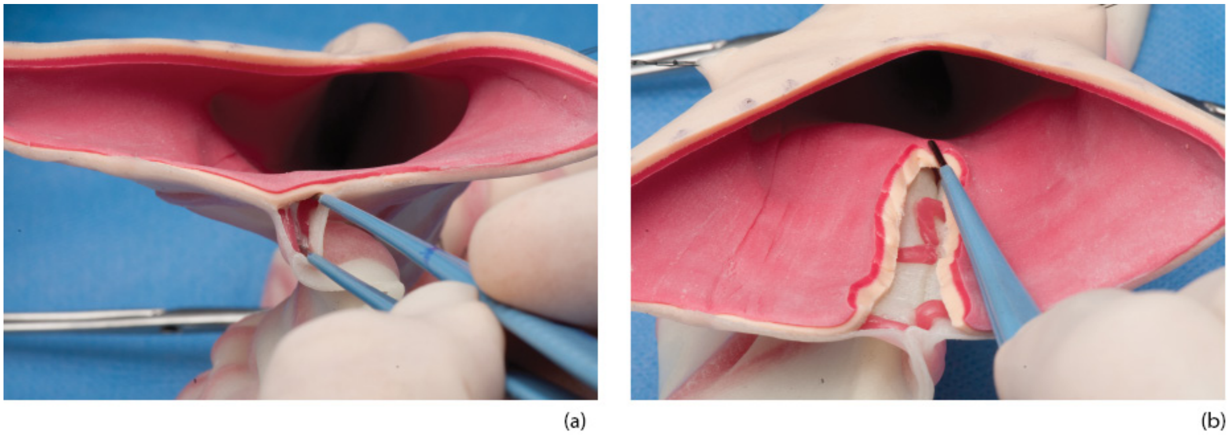
52.3

The dilated segment is divided in a distal to proximal direction. The antimesenteric border is than cut longitudinally using bipolar forceps passing between the traction sutures for a comfortable working length of about 2–5 cm (**Figure 52.4**).



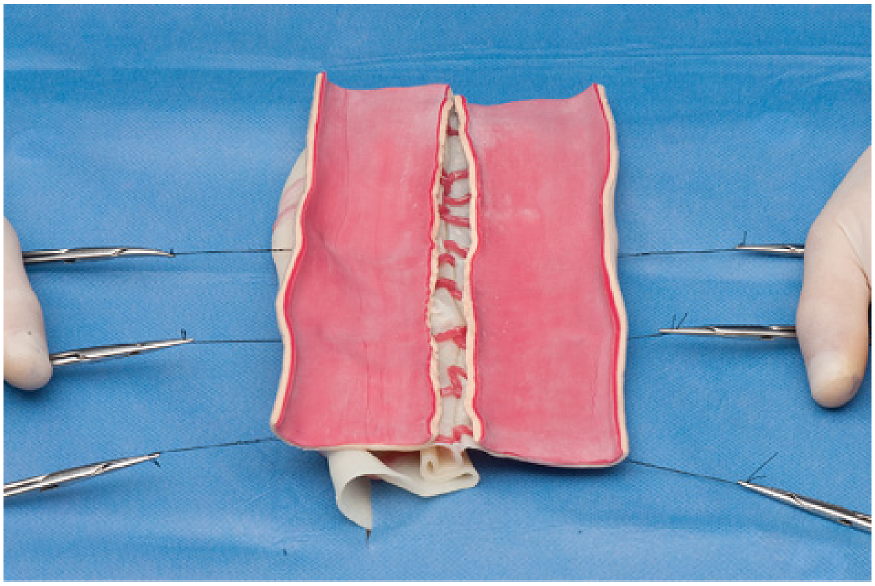
52.4

Using blunt/bloodless dissection, the two mesenteric leaves are separated creating an idealized triangle (**Figure 52.5a**). The mesenteric border is divided longitudinally passing between the blood vessels using the cutting mode of the bipolar forceps (**Figure 52.5b**).


(a) (b)

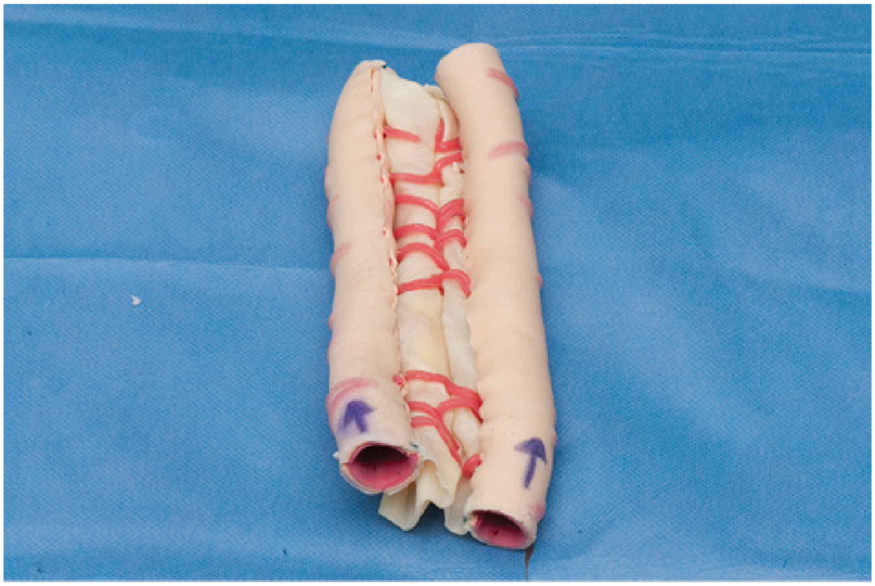
52.5a,b

These stages are then repeated moving proximally until the dilated bowel is divided into two fully vascularized hemiloops (**Figure 52.6**).



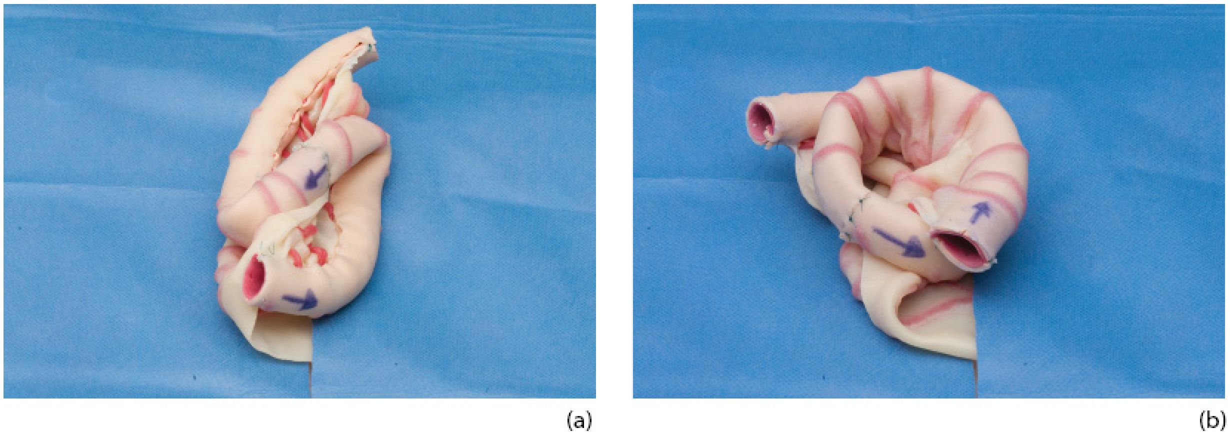
52.6

One hemiloop is completely detached from the bowel by division along the lateral wall. The other hemiloop is now tubularized, starting proximally in continuity with the duodenum. We use a continuous horizontal inverting mattress suture (5/0 Maxon□) with locking knots every three throws. Sutures are placed 2 mm away from the cut edge of the bowel, turning the edges into the lumen. It is important to avoid injury to the blood supply arising in the mesenteric aspect. The second hemiloop is then tubularized and anastomosed isoperistaltically to the first (**Figure 52.7**).



52.7

Isoperistaltic anastomosis requires apposition of opposite ends of the hemiloops. This can be performed in the shape of a “Bianchi–S” with the bowel lying over the mesentery (**Figure 52.8a**), or as an “aigrain spiral” with one loop passing beneath the other (**Figure 52.8b**). The distal end of the second hemiloop is anastomosed to the distal bowel (often the colon) to establish bowel continuity.



52.8a,b

POSTOPERATIVE CARE

Intensive care management is appropriate for 24–48 hours. Intestinal function usually restarts within 5–6 days and enteral feeding can then be slowly re-established. It is not unusual for patients to experience abdominal cramps or vomiting immediately during the postoperative time as a result of initial delay in the passage of food through the lengthened segment.

COMPLICATIONS

No operative mortality has been reported in the authors' experience so far. LILT-related complications reported in the literature are anastomotic leak, stenosis, external fistula formation from suture line disruption, and very rarely ischemic loss of one of the hemiloops. D-lactic acidosis is an uncommon complication related to excess of production of D-lactate by intestinal bacteria due to incomplete absorption of carbohydrate. In our experience, this event was reported in only eight patients and it resolved with dietary manipulation associated with a carbohydrate-restricted diet, no absorbable antibiotics, and oral bicarbonate. Severe stenosis or sepsis due to significant dilatations may warrant further surgery. Bowel-lengthening procedures have the potential for serious complications and should not be undertaken lightly or in a center with little experience.

OUTCOME

Data from the literature suggest a different degree in enteral autonomy success after LILT as shown in Table 52.1. The authors' personal experience is of 63 LILT procedures without operative mortality. Full enteral autonomy in this cohort was achieved in 92% of cases within 18 months after LILT. Tailoring and lengthening aim to reduce the amount of PN providing a quality of life improvement but, unfortunately, no recent data have been reported in the literature. Interestingly, a recent review has shown that the survival rate after an LILT procedure was 89% in the last 15 years.

Table 52.1 Long-term outcomes after LILT

		Small bowel length (cm)					
Author (year)	**N**	**Pre-LILT**	**Post-LILT**	**Follow-up (years)**	**Transplant-free survival (%)**	**Enteral autonomy (%)**	**Reporting center**
Khalil et al. (2012)	19	60 (18–140)	90 (37–260)	n/a	92	91	Manchester, UK
Reinshagen et al. (2008)	53	30.6 (±15.1)	54.71 (±26.95)	6.6 (0.5–19.5)	77.36	75.86	Munich, Germany
Hosie et al. (2006)	49	27 (12–60)	51 (18–120)	n/a	81.63	47.5	Heidelberg, Germany
Bueno et al. (2001)	26	28 (8–70)	48 (16–100)	2.4 (0.29–11.2)	53.84	n/a	Pittsburgh, USA
Bianchi (1999)	20	42.5 (25–110)	68 (50–166)	6.8 (1–15.5)	n/a	77.7	Manchester, UK

The success of LILT seems to be strictly related to the correct selection of patients. According to some authors, the LILT procedure is a challenging technique compared to other intestinal rehabilitation procedures. Data from the literature have shown that the presence of a dilated bowel segment (>4 cm) and no fibrotic mesentery seem to be the two mandatory inclusion criteria in order to select the right patient for the LILT procedure.

CONCLUSION

LILT is to be considered the first described effective procedure that changes dramatically the outcome for the short bowel patients, setting the scene for the modern treatment of this severe condition. In patients where bowel length is critical (<20 cm), the LILT operation accomplishes intestinal tapering without loss of surface area. LILT has the ability to recreate a physiological segment of bowel, to improve intestinal motility, and to increase mucosal surface area. LILT should be performed with extreme caution in patients with ultra-short bowel and in the presence of liver disease.

SERIAL TRANSVERSE ENTEROPLASTY (STEP)

PRINCIPLES AND JUSTIFICATION

In serial transverse enteroplasty (STEP), a stapler is placed perpendicularly to the mesentery and applied at intervals from opposite sides to create a zigzag channel. The surgeon determines bowel caliber by the position of the staple lines. In contrast to LILT, which decreases the bowel circumference by half regardless of the degree of dilation, STEP facilitates autologous reconstruction in patients with varying degrees of bowel dilation. Additional advantages of STEP include the absence of intestinal anastomoses, reduced potential for vascular compromise, and decreased technical complexity when compared with other bowel-lengthening procedures.

SBS with bowel dilation and failure to progress toward enteral autonomy remains the most common indication for STEP. It significantly increases bowel length and absorptive capacity, with reported gains of up to double the bowel length following the procedure. Additionally, refractory bacterial overgrowth is an indication for STEP. By tapering dilated bowel, STEP can reduce bacterial overgrowth and improve motility. In neonates with marginal bowel length and significantly dilated proximal bowel due to atresia, a primary STEP procedure can also be considered.

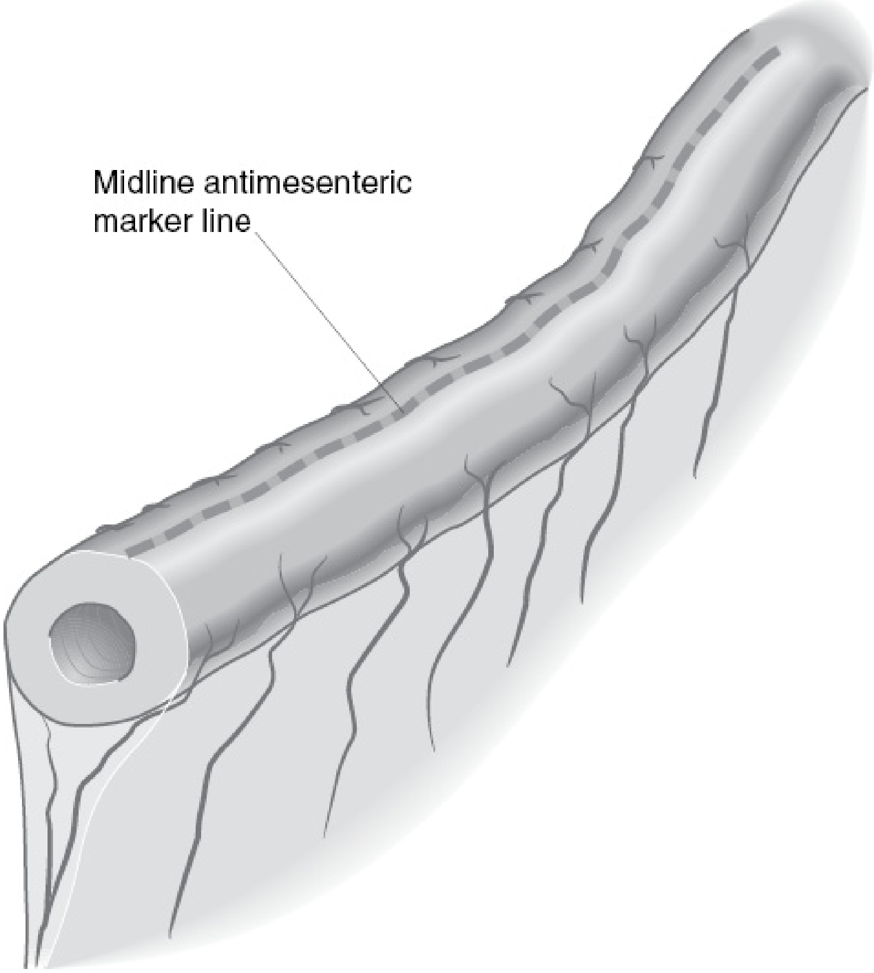
We hypothesize that tapering and lengthening of the previously dilated bowel facilitates gut adaptation and allows enterocyte mass to increase. This is supported by the demonstration of increased levels of citrulline, a non-protein amino acid that correlates with enterocyte mass, in both animals and humans following STEP. In addition, increased levels of the gut growth factor GLP-2 have been noted in rodents which have undergone a modified STEP operation. Small bowel motility in porcine models of SBS was preserved following STEP, when compared with control animals. Nevertheless, the additional length gained after bowel-lengthening procedures should not be considered necessarily equivalent to a similar length of native healthy bowel.

PREOPERATIVE INVESTIGATIONS

As with the LILT procedure, a trial of medical and nutritional therapy such as enteral feeding to promote bowel adaptation, post-pyloric feeding, motility agents, and culture-directed antimicrobial therapy for bacterial overgrowth should be considered before proceeding with STEP. Mechanical obstruction leading to enteral intolerance should be ruled out. All patients should receive an upper gastrointestinal series to assess degree of bowel dilation and guide surgical decision making.

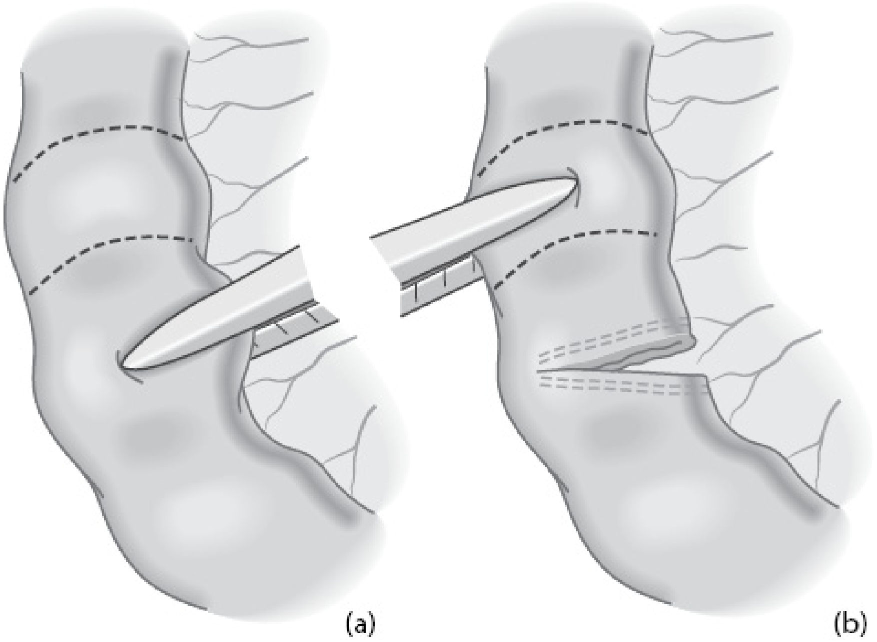
OPERATION

Laparotomy may be performed through an existing surgical incision, or through a transverse or midline exposure. The entire small bowel is freed of adhesions to define all areas of dilation. The dilated bowel is measured and marked along the antimesenteric border to facilitate the perpendicular application of the stapler to the mesentery (**Figure 52.9**). The distal bowel is addressed first in the procedure, and then proximal progress is made.



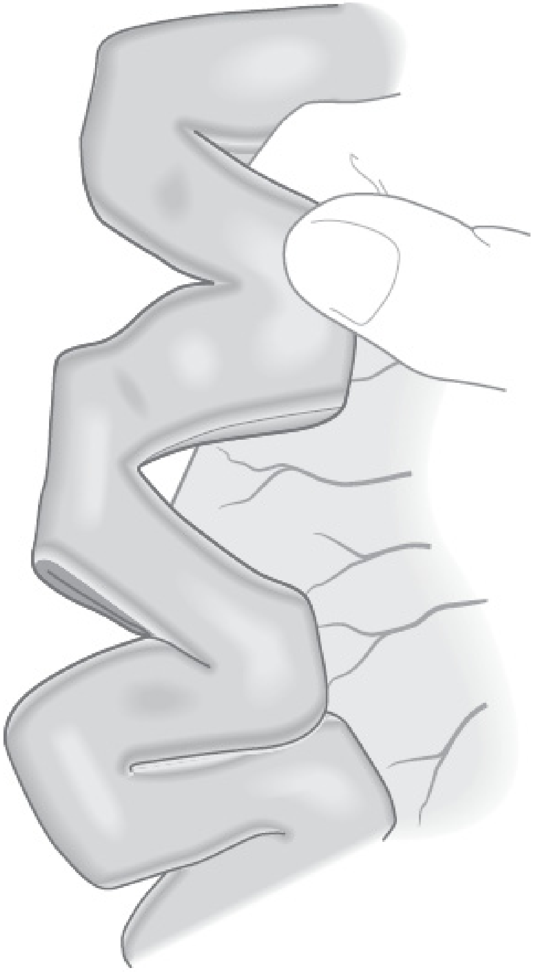
52.9

A window is created in the mesentery to accommodate passage of a GIA stapler (**Figure 52.10a**). In larger children and adults, a GIA stapler is utilized. In neonates and smaller children, the Endo GIA□ is preferred. The stapler is inserted through the window at a 90-degree angle to the mesentery and fired. A size 18 red rubber catheter can be placed on the lower anvil of the stapler to guide it through the defect and reduce the chance of vascular injury. The degree to which each staple line crosses the mesentery is determined by the surgeon, according to the desired channel width and extent of bowel dilatation (**Figure 52.10b**). The non-dilated duodenum often provides a guide for the appropriate diameter. In neonates, the STEP channel is usually 1.5–2 cm.



52.10a,b

The process of creating a mesenteric defect, passing, and firing the stapler is repeated at intervals throughout the length of the dilated bowel. Each staple line approaches the bowel from the opposite direction of the previous staple firing, creating a zigzag channel (**Figure 52.11**). The distance between staple lines is again affected by bowel dilation and desired channel width, and it varies based on age and caliber of normal bowel. As the bowel becomes less dilated, there is decreased overlap of staple firings. A figure-of-eight PDS® suture is placed at the staple line apex to decrease the risk of leak from the weakest area.



52.11

COMPLICATIONS

These are similar to those after LILT but also include recurrent dilation, staple line leaks, and gastrointestinal bleeding due to ulcers along the staple line. Although there are no anastomoses with the STEP, bowel obstruction and prolonged ileus can occur. Sepsis, abscess formation, and catheter-related bacteremia have also been described. For those presenting with recurrent dilation, repeat STEP has been shown to be a viable option for management, with progression to enteral autonomy after the repeat STEP reaching over 40% in one series.

A video of the STEP is available online: https://www.youtube.com/watch?v=jl7AjiyXwQ&t=824s

OUTCOME: STEP DATA REGISTRY

The web-based International Serial Transverse Enteroplasty Data Registry for patients was established in 2004 (https://apps.childrenshospital.org/externalClinical/STEP/). Jones et al. summarized the largest experience to date in 111 patients from 50 centers in 14 countries. The most common underlying disease process was gastroschisis, with or without concomitant volvulus (45%), followed by intestinal atresia (34%) and NEC (8%). Of the 97 registry patients with adequate follow-up following first STEP, 87 (89%) were transplant-free survivors. Over half of these patients achieved enteral autonomy and overall two-thirds had improved enteral tolerance. A subgroup analysis of patients undergoing STEP for SBS with PN dependence identified longer small bowel length as the most significant preoperative factor associated with attainment of enteral autonomy following STEP. Median time to reach enteral autonomy in the entire cohort was 21 (95% CI: 12–30) months.

As with LILT, deaths following STEP were secondary to concomitant IFALD and sepsis. Multivariate analysis of the entire cohort identified significantly higher preoperative serum direct bilirubin and significantly shorter preoperative bowel length in those who progressed to transplantation or death, when compared with transplant-free survivors. These factors, along with the median time needed to wean from PN, must be weighed when considering a candidate for STEP. Patients with severe complications of short bowel syndrome and IFALD including severe bleeding, loss of access, and end-stage liver disease should be considered for small bowel transplantation in lieu of STEP or other bowel lengthening procedures.

Table 52.2 illustrates these outcomes and demonstrates that enteral autonomy may be achieved in anywhere between 24% and 60% of patients following STEP. Small bowel length, a marker for achieving enteral autonomy, is also significantly greater post-STEP. While the definition of success has focused on the ability to achieve enteral autonomy, modest but important benefits can be identified following STEP. Decreasing PN dependence substantially improves a child's quality of life, improves IFALD, decreases the burden of short bowel syndrome on the family, and should be accounted for when considering STEP. One center recently reported a 40% median increase in enteral tolerance just 6 months following surgery, while another series accounted for a notable decline in median PN calories at 1-year follow-up. Long-term studies comparing outcomes after STEP versus contemporary non-surgical rehabilitation efforts are still lacking. As fluid and macronutrient requirements per kilogram of body weight for growth and development decline with age, the decrease in PN dependence may not be fully attributed to the bowel-lengthening procedure.

Table 52.2 **Long-term outcomes after STEP**

Author (year)	*N*	Small bowel length (cm) Pre-STEP	 Post-STEP	Follow-up (years)	Transplant-free survival (%)	Enteral autonomy (%)	Reporting center
Barrett et al. (2016)	17	55.6 (±27.5)	Original "+19.5 cm"	3 (1.5–10)	94	24	Ann Arbor, MI, USA
Wester et al. (2016)	27	54 (15–93)	71 (27–150)	3.8 (0.2–10.5)	96	59	Sweden and Finland
Mercer et al. (2013)	51	51 (15–175)	87 (31–270)	3.25 (0.5–7)	82	48	Omaha, NE, USA
Javid et al. (2013)	16	84 (19–295)	103 (24–375)	2.2 (0.1–4.6)	75	60	Seattle, WA, USA
Ching et al. (2009)	16	60 (IQR 30, 108)	117 (IQR 62, 209)	1.9 (0.1–5.9)	88	40	Boston, MA, USA

IQR, interquartile range.

CONCLUSION

STEP has proven successful in increasing enteral tolerance in patients with intestinal failure through mechanisms thought to involve enhanced motility, reduced bacterial overgrowth, increased enterocyte mass, and improved nutrient absorption. It has been shown to have broad applicability and remains the preferred management at many centers for surgical reconstruction due to its technical simplicity and safety. As in LILT, progression of IFALD and sepsis remain the major sources of morbidity and mortality in patients undergoing STEP.

VIDEOS

OPS_8e_video_52.1 Longitudinal Intestinal Lengthening and Tailoring (LILT) procedure

OPS_8e_PPT_52.2 Serial transverse enteroplasty (STEP): Boston Children's Hospital https://www.youtube.com/watch?v=Zjl7AjiyXwQ&t=824s

FURTHER READING

Barrett M, Demehri FR, Ives GC, et al. Taking a STEP back: Assessing the outcomes of multiple STEP procedures. *J Pediatr Surg* 2017; 52: 69–73.

Bianchi A. Intestinal loop lengthening – a technique for increasing small intestinal length. *J Pediatr Surg* 1980; 15: 145–51.

Ching YA, Fitzgibbons S, Valim C, et al. Long-term nutritional and clinical outcomes after serial transverse enteroplasty at a single institution. *J Pediatr Surg* 2009; 44: 939–43.

Coletta R, Khalil BA, Morabito A. Short bowel syndrome in children: Surgical and medical perspectives. *Semin Pediatr Surg* 2014; 23: 291–7.

Duggan CP, Jaksic T. Pediatric intestinal failure. *N Engl J Med* 2017; 377: 666–75.

Garnett GM, Kang KH, Jaksic T, et al. First STEPs: Serial Transverse Enteroplasty as a primary procedure in neonates with congenital short bowel. *J Pediatr Surg* 2014; 49: 104–7.

Javid PJ, Sanchez SE, Horslen SP, Healey PJ. Intestinal lengthening and nutritional outcomes in children with short bowel syndrome. *Am J Surg* 2013; 205: 576–80.

Jones BA, Hull MA, Potanos KM, et al. Report of 111 consecutive patients enrolled in the International Serial Transverse Enteroplasty (STEP) Data Registry: A retrospective observational study. *J Am Coll Surg* 2013; 216: 438–46.

Kim HB, Fauza D, Garza J, et al. Serial transverse enteroplasty (STEP): A novel bowel lengthening procedure. *J Pediatr Surg* 2003; 38: 425–9.

Mercer DF, Hobson BD, Gerhardt BK, et al. Serial transverse enteroplasty allows children with short bowel to wean from parenteral nutrition. *J Pediatr* 2014; 164: 94–8.

Murphy F, Khalil BA, Gozzini S, et al. Controlled tissue expansion in the initial management of the short bowel state. *World J Surg* 2011; 35: 1142–5.

Pakarinen MP, Kurvinen A, Koivusalo AI, et al. Long-term controlled outcomes after autologous intestinal reconstruction surgery in treatment of severe short bowel syndrome. *J Pediatr Surg* 2013; 48: 339–44.

Wester T, Lilja HE, Stenstrom P, Pakarinen MP. Absent ileocecal valve predicts the need for repeated step in children. *Surgery* 2016; 161: 818–22.

53

Surgery of portal hypertension

CAROLINE LEMOINE AND RICCARDO A. SUPERINA

HISTORY

The use of surgery as a tool to treat the symptoms of portal hypertension (PHT) is relatively recent. Nicholas Eck, a Russian surgeon, described an experimental end-to-side portocaval shunt in dogs toward the end of the 19th century to show that some could survive it. This principle was first applied in humans in the 1950s to alleviate PHT though profound encephalopathy limited its application. More peripheral portosystemic shunts which aimed to preserve hepatopetal portal venous flow followed, such as the distal splenorenal (DSR) shunt (Warren procedure) in the 1960s and 1970s. In addition, De Ville de Goyet and colleagues from Louvain in Belgium described a more physiologically beneficial solution, the surgical meso-Rex bypass (MRB), for an obstructed portal vein post-transplant and then this technique was utilized in children with idiopathic non-transplant portal vein obstruction. Although the pathophysiology and principles of management of PHT have been well described in adults, and only later applied to the treatment of children, the causes of PHT and the hemodynamic consequences are clearly different, and the options for treatment and recommendations therefore, are different.

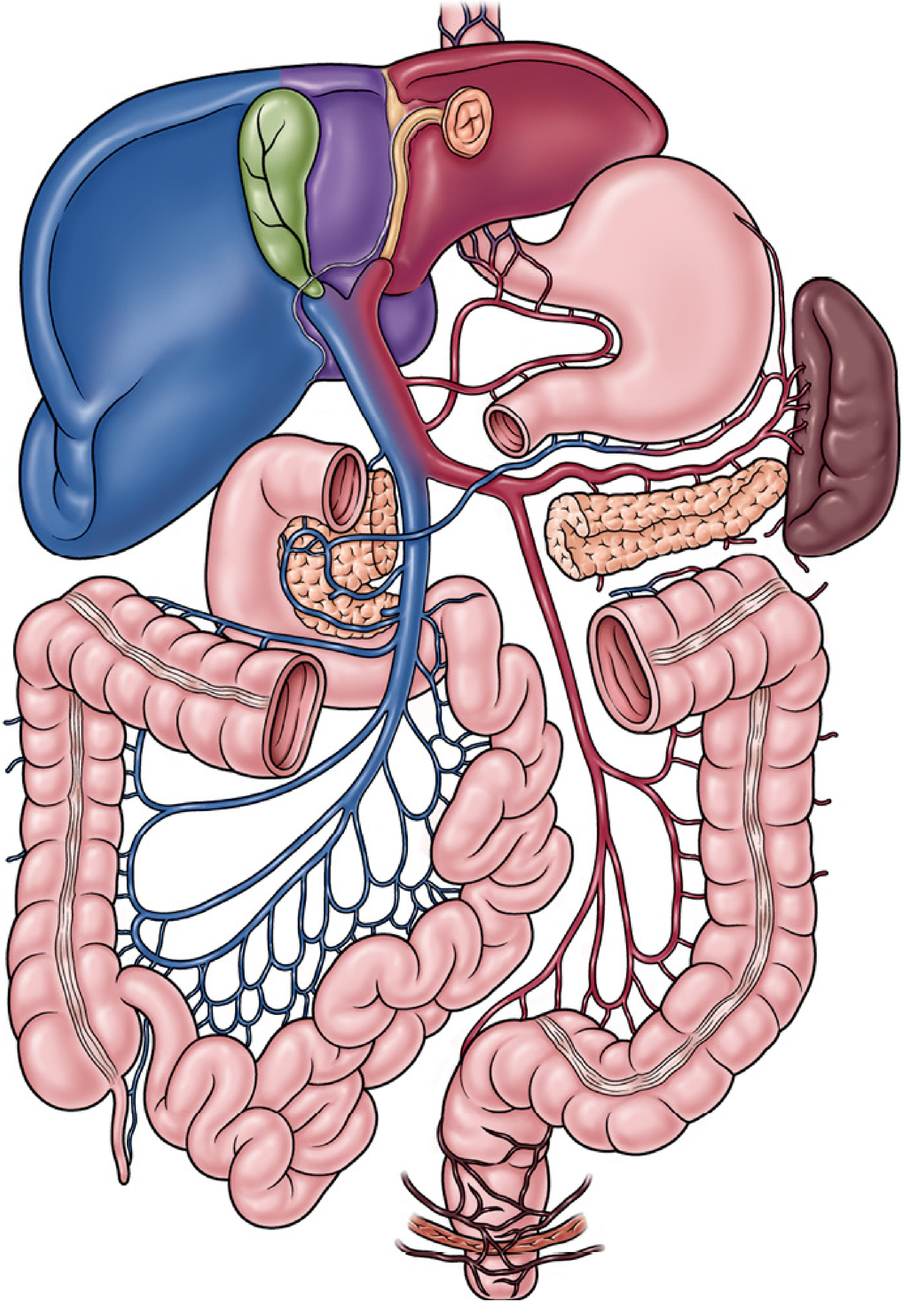
PRINCIPLES AND JUSTIFICATION

Surgical shunts are now rarely used in adults. However, they remain a valuable option in the long-term management of PHT in children. The MRB, for those with extrahepatic portal vein occlusion (EHPVO), effectively cures PHT and minimizes or even eliminates its major symptoms by restoring physiologic blood flow to the liver. The introduction of this procedure has created a major shift in the timing of intervention for those patients.

Portosystemic shunts (PSSs) such as the distal splenorenal shunt (DSRS) and the mesocaval shunt (MCS) can also be valuable tools to be used in selected situations in stable children who have no clinically significant liver disease such as those children with EHPVO who are not MRB candidates. Selective PSS can also be applied to those patients whose liver disease has not progressed or is not expected to progress to the point where transplantation may be necessary. PSS can be used to maintain quality of life, avoid repeated general anesthesia for esophagoscopy for banding or sclerotherapy and sometimes be all that is necessary to treat a child with PHT.

Definitions

PHT occurs when portal pressure is >10 mmHg or when hepatic venous pressure gradient (HPVG) is >5 mmHg. In the adult population, it has been shown that, when HPVG reaches 12 mmHg, patients are at risk for suffering from a variceal bleed, most often from esophageal varices (**Figure 53.1**).



53.1

PHT can also be defined clinically as an increase in portal and mesenteric venous pressure that results in the development of:

- Alternative venous collateral channels, e.g. varices
- See **Figure 53.1**
- Hypersplenism, e.g. splenomegaly with associated thrombocytopenia
- Other clinical manifestations including ascites, spider nevi, *caput medusae* and gastrointestinal bleeding

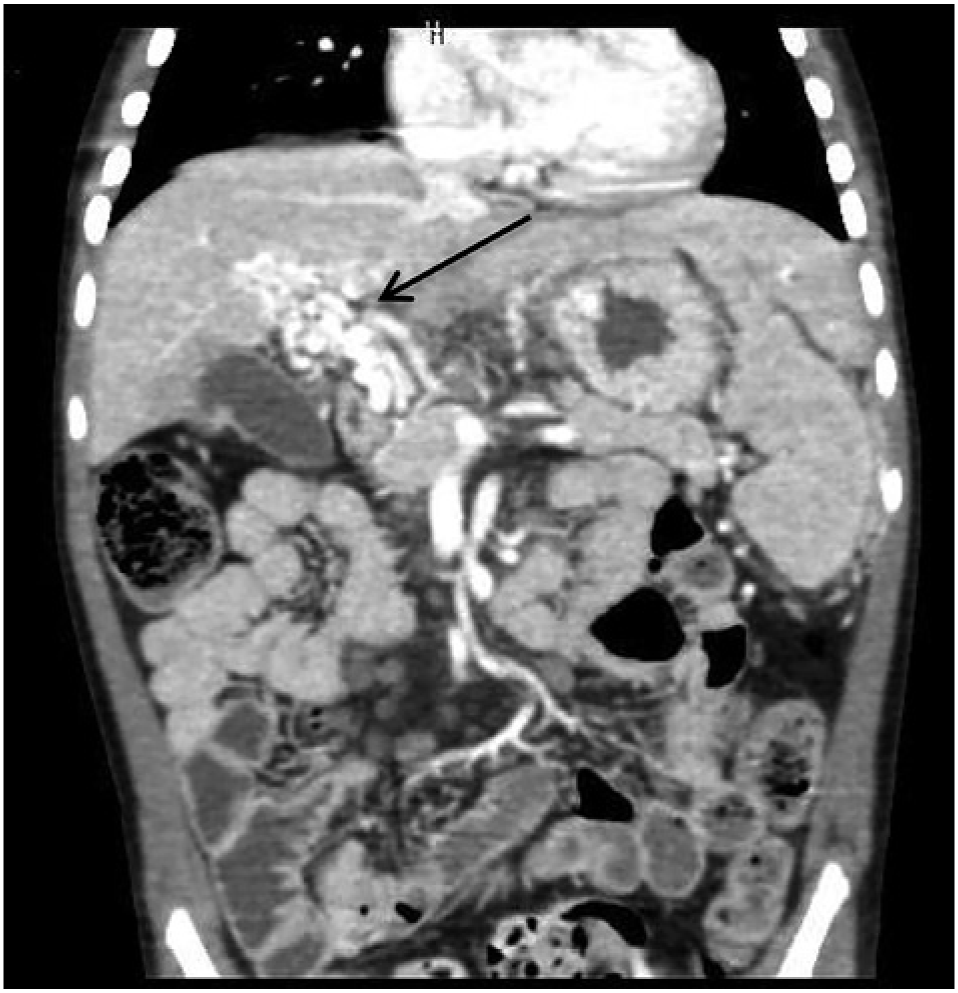
Classification

PHT can be subdivided in prehepatic, hepatic, and posthepatic causes (**Table 53.1**) depending on where the greatest impediment to portomesenteric venous flow is felt to be. Differentiating between cirrhotic and non-cirrhotic PHT is essential in terms of management and those with underlying decompensated cirrhosis should be referred for liver transplantation (LT) rather than surgical shunting.

Table 53.1 **Classification of PHT**

Prehepatic	EHPVO	Congenital Neonatal acquired (e.g. umbilical vein catheter) Childhood sepsis Tumor Trauma Hypercoagulable condition
Hepatic	Cirrhotic	Biliary atresia Alagille syndrome PFIC Cystic fibrosis Alpha-1-antitrypsin deficiency Autoimmune disease
	Non-cirrhotic	Congenital hepatic fibrosis Post-schistosomiasis
Posthepatic		Budd–Chiari syndrome Veno-occlusive disease Pericarditis Right-sided heart failure

Chief among the prehepatic causes of PHT is EHPVO, which is reported to cause up to 50% of cases in the pediatric population (**Figure 53.2**). Risk factors for EHPVO include history of umbilical vein catheter placement, omphalitis, and a hypercoagulable state. This can be successfully treated with a MRB in up to 70–80% of cases. Other surgical alternatives in children with EHPVO who are anatomically unsuitable include MCS shunt.



53.2 CT scan of a patient with EHPVO. Note the presence of a cavernoma in the hilum of the liver (black arrow).

CLINICAL PRESENTATION

Patients with PHT may present with a wide variety of initial symptoms, depending on the underlying diagnosis. Some may be entirely asymptomatic until the first presentation with an upper gastrointestinal (GI) hemorrhage (hematemesis and/or melena). Others may be diagnosed with idiopathic splenomegaly and undergo an oncologic workup only to be found later to have PHT. Others may have known liver disease before the onset of PHT.

Patients with asymptomatic EHPVO should be referred early for surgical evaluation. The MRB can now be considered as "pre-primary prophylaxis" (intervening before the occurrence of a first GI bleeding episode). However, surgery is rarely if ever needed in the acute presentation and careful imaging and investigation should be performed once the patient has stabilized.

Referral to a specialized center with both PHT surgery and transplant expertise is recommended. Surgical success depends on factors including volume of cases, and expertise in liver and vascular surgery of the surgeon. A child's chance at fully benefiting from a surgical shunt or bypass is at the time of the initial surgery. Previous failed surgical attempts by less experienced surgeons will unfortunately usually adversely affect the results at subsequent operations when the child is finally referred to a center of excellence.

MANAGEMENT OF PHT: DIFFERENCES BETWEEN ADULTS AND CHILDREN

In adults, primary (prevention of a first variceal bleed) and secondary prophylaxis (prevention of rebleeding) includes β-blocker medication and serial endoscopic variceal banding. β-blockers act through several mechanisms of action: By reducing cardiac output (β1-receptor antagonism); induction of splanchnic vasoconstriction and therefore reduction of portal venous flow (α-receptor); and through an antagonist effect of norepinephrine, they induce the constriction of intrahepatic myofibroblasts, activated stellate cells, and vascular smooth muscle cells. However, the use of β-blockers in children remains currently not proven. Beta blockade prevents tachycardia, the natural hemodynamic response to bleeding in children and also lowers the systemic blood pressure, which is deleterious in the setting of hypovolemic hemorrhagic shock.

In children, upper GI endoscopy for variceal banding is performed under general anesthesia. The long-term sequelae of repeated general anesthesia events on the neurological development of children remain unknown but there is some evidence to suggest that there may be a cumulative effect that impacts normal development in the very young.

Variceal banding is technically not possible in patients weighing <10 kg because of the size of the endoscope. Sclerotherapy is then used, but this is associated with risks of esophageal stricture and perforation. Of course, local control of varices does not solve the underlying cause of PHT and does not treat hypersplenism. Early surgical intervention is recommended for the management of PHT in the children.

We do not feel that splenectomy by itself has a role in the management of PHT in children and preservation should always be a goal in whatever operation is chosen. Splenectomy as an isolated procedure performed with the goal of relieving hypersplenism will always be successful in restoring the platelet count back to normal but this will come at a price. The PHT itself will remain unchanged and variceal hemorrhage may actually be exacerbated by eliminating the spleen as a vascular reservoir.

Transjugular intrahepatic portosystemic shunts (TIPS) are commonly used in the management of PHT in adults patients. Most reports of TIPS placement in pediatric patients are in teenagers. Although technically feasible, TIPS placement in children has a variable success rate, and is associated with high re-intervention rates and up to 40% rebleeding occurrence post-procedure. TIPS placement in those patients <10 kg has been associated with a high incidence of complications (death and thrombosis). Therefore, in our practice, TIPS placement is limited to a bridge to liver transplant in the setting of intractable variceal bleeding refractory to endoscopic and medical interventions.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

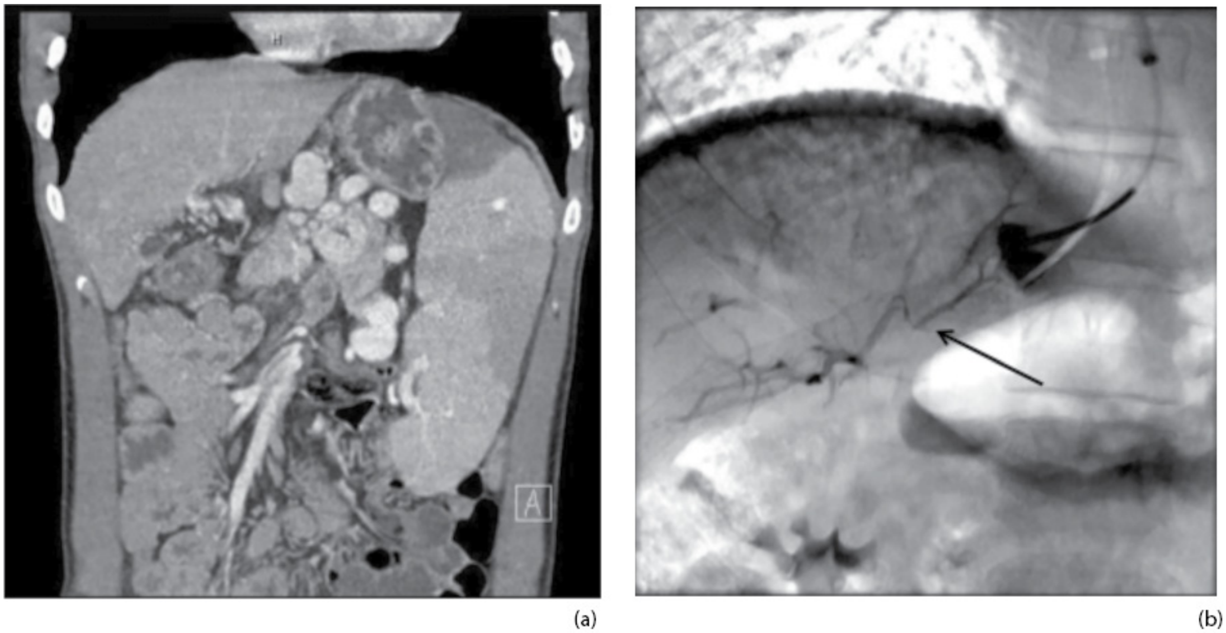
The evaluation of a child with PHT starts with laboratory studies that assess the degree of hypersplenism and liver dysfunction. These include a complete blood count (CBC), a comprehensive metabolic panel including liver function tests (including albumin and bilirubin), serum ammonia, and a coagulation panel. These should be obtained to make sure there is little evidence of impaired liver function that would lead to referring the patient for a transplant evaluation.

In cases of EHPVO, an extensive hypercoagulability workup is performed to exclude a prothrombotic state as the cause of the initial thrombosis. This workup should include the following investigations: Genetic mutational analysis for Factor V Leiden, methylenetetrahydrofolate reductase (MTHFR), and prothrombin; and assays for protein C and S, antithrombin III levels, and a complete panel of antiphospholipid antibodies.

Abdominal ultrasound (US) with Doppler flow studies of the liver is often obtained but is of very limited usefulness in determining what kind of surgery is possible. A US will generally confirm the diagnosis of a portal cavernoma. An abdominal computed tomography (CT) scan with portal venous phase contrast should be obtained to evaluate the patency of the superior mesenteric vein (SMV), splenic vein, renal vein, and inferior vena cava (IVC). This provides extensive information to the surgeon on vascular anatomy, and on which type of shunting procedure can be offered to the patient. CT imaging may also help identify other possible venous conduits to be used in the bypass operation, including the inferior mesenteric vein (IMV) or coronary (left gastric) vein.

A bilateral neck Doppler US is performed preoperatively to evaluate patency and size of both internal jugular veins (IJVs) before as part of the preoperative work up for an MRB.

However, preoperative CT scans will often fail to show anything of the patency of the intrahepatic portal venous circulation and patients should not be ruled out as candidates for MRB solely on such cross-sectional imaging (**Figure 53.3a**). A retrograde transjugular venogram with hepatic vein pressure measurements should be performed in order to show a patent and suitable intrahepatic portal vein that will serve as a landing spot (**Figure 53.3b**). An increased hepatic vein pressure gradient may reflect abnormalities in the liver parenchyma that would caution the surgeon from going ahead with an MRB. A liver biopsy is always done in order to complete the assessment of the liver.



53.3a,b (a) Preoperative CT of a patient with EHPVO. Note the absence of intrahepatic portal venous branches on CT (portal venous phase); also note the size inversion between the liver (smaller) and the spleen (larger). (b) Preoperative venogram, same patient: Demonstrates a patent intrahepatic portal vein (black arrow), with communication between the left and right intrahepatic portal venous branches.

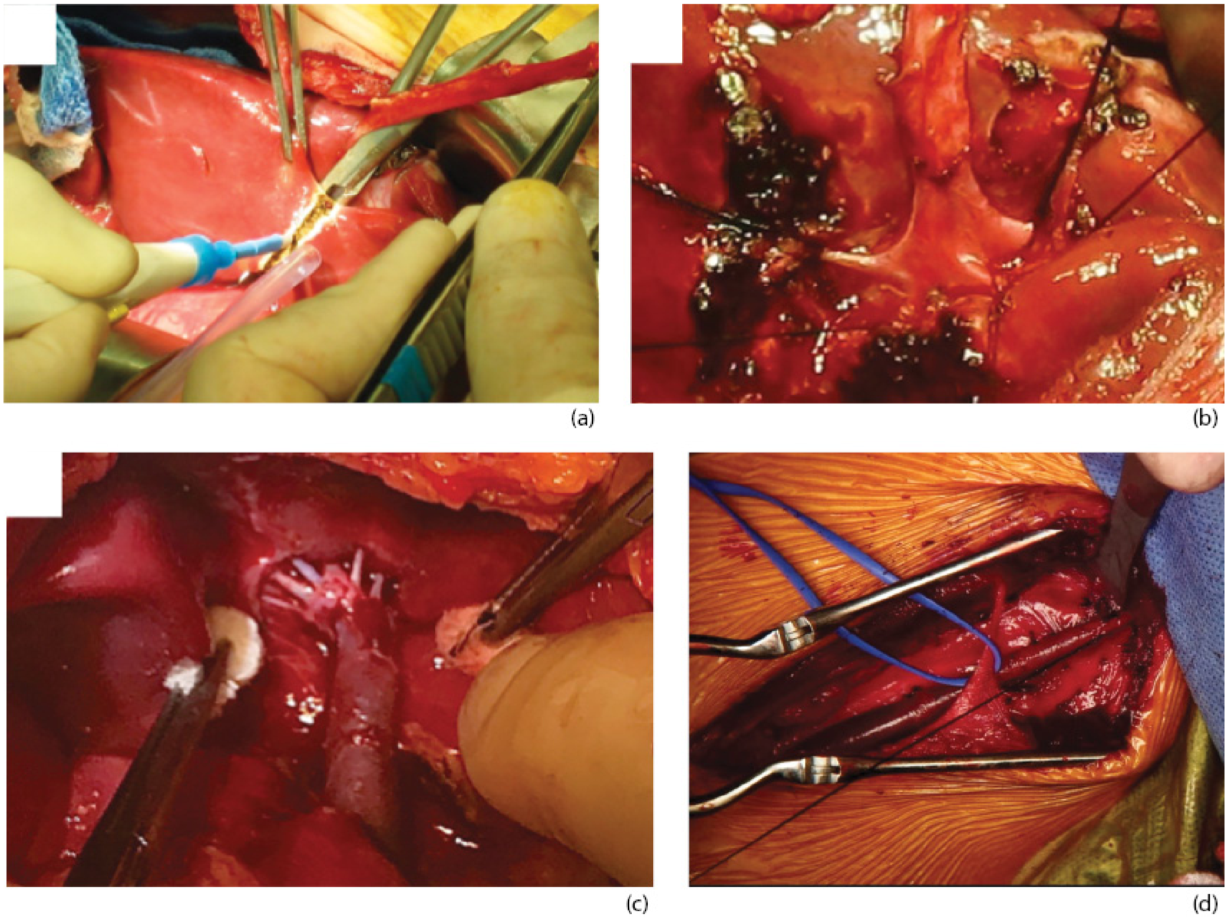
The preoperative workup for MRB also includes a bilateral jugular vein Doppler US to ensure patency of both veins.

Patients with long-standing portosystemic shunting from PVT are at risk for developing pulmonary hypertension. Vasoactive substances from the mesenteric circulation are normally cleared by the liver before reaching the pulmonary circulation. In PHT, they bypass the liver through the spontaneous portosystemic shunts and collaterals, and cause progressive vasoconstriction of the pulmonary vasculature. Therefore, an echocardiogram must also be performed preoperatively. A patient who is found to have pulmonary hypertension by echo requires extensive cardiological evaluation including possible cardiac catheterization. These patients are at high risk for suffering from a pulmonary hypertensive crisis postoperatively, which can be fatal. A patient with either pulmonary hypertension or hepatopulmonary syndrome, who is not an MRB candidate based on preoperative investigation, should be evaluated for a liver transplant. Shunts such as a DSRS or an MCS are not advisable in these patients since the shunting will only increase after the operation and exacerbate existing portopulmonary or hepatopulmonary syndromes.

OPERATIONS

Meso-Rex bypass

The meso-Rex bypass (MRB) (**Figure 53.4a–d**) is the preferred surgery for the management of EHPVO, in the absence of intrinsic liver disease. The routine use of a preoperative venogram has helped better identify which patients will successfully undergo an MRB. Nevertheless, patients and their family are informed that preoperative imaging may not always be accurate and that, in the operating room, an MRB may prove to be impossible to do successfully despite favorable preoperative imaging. In this event, a DSRS is then performed during the same surgery, as it effectively corrects hypersplenism, and reduces the incidence of GI bleeding. This approach, however, is not suitable for the asymptomatic patient, who would not benefit from a prophylactic MRB. In these patients, the surgeon must be as certain as possible that the MRB will be possible before embarking on the operation.



53.4a–d (a) The Rex recessus is accessed by dividing the bridge of liver between segments 2–3 and segment 4. (b) The portal venous branches draining in the intrahepatic portal vein are dissected; they have been isolated and encircled with Pott's ties. (c) The patient's head is rotated to the right (draped under the surgical drapes to the right of the picture, the patient's chest is to the left of the picture). This photograph shows the left IJV that has been dissected on its entire length, from its insertion in the subclavian vein up to the base of the skull at the jugular foramen. All its branches, including the facial vein, have been ligated. Note that the external jugular vein has been preserved (encircled by the blue vessel loop) to avoid postoperative neck and face swelling. (d) Distal end of the MRB, showing the various portal venous branches.

1. Incision options include either a midline longitudinal incision or a limited transverse subcostal incision with a vertical extension. Both offer good surgical exposure, and the decision to perform one or the other is based on the physiognomic characteristics of the child.
2. On entering the abdominal cavity, the round ligament is identified and ligated. The falciform ligament is taken down until reaching the level of the hepatic veins. The Rex recessus is often located behind a bridge of

liver parenchyma extending between segments 3 and 4. This bridge needs to be divided and often portions of the liver excised to gain access to the insertion of the round ligament in the intrahepatic left portal vein, and also to create room for the future vein graft, avoiding any external compression.

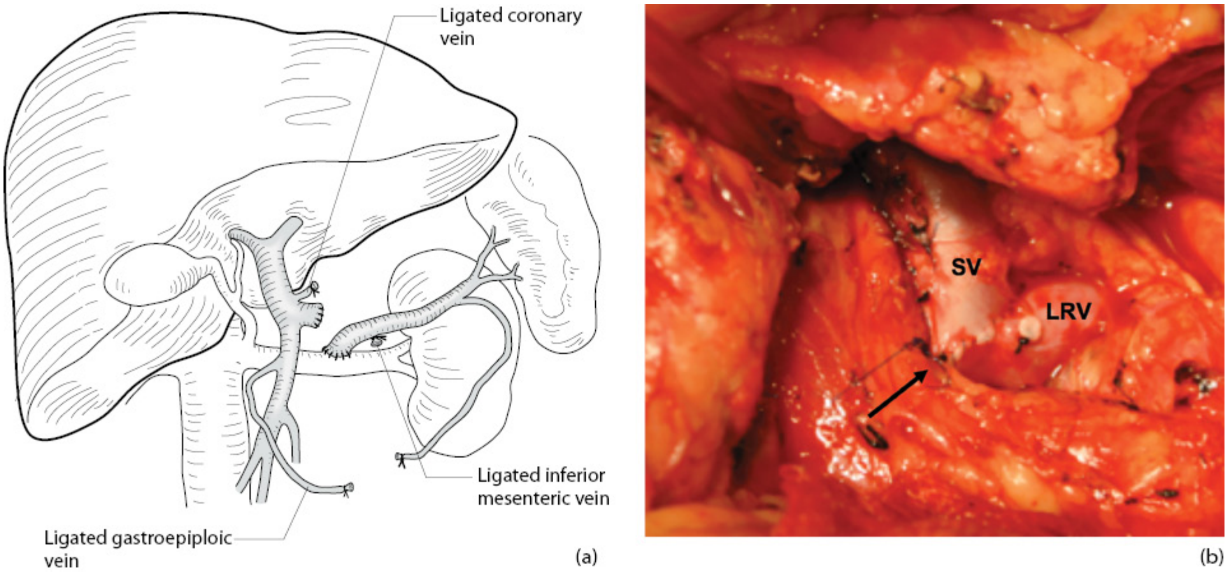
3. The patient is then placed in Trendelenburg position. The peritoneum covering the round ligament is divided. All portal venous branches extending in the liver parenchyma are encircled with Pott's silk ties, taking care not to ligate them. Once the intrahepatic left portal vein is reached, the patient is fully heparinized (100 units/kg). The portal venous branches are encircled twice, while the main intrahepatic portal vein confluence is clamped using a fine vascular clamp. The portal vein should be opened to assess the patency of its lumen (which should be lined with venous endothelium) and confirm quality of back bleeding. If the vein is found to be atretic or fibrotic (fibrous "spider web" appearance), or if no patent lumen or good venous backflow can be identified, then a DSRS should be performed instead as the chances of maintaining a good enough outflow network to keep the MRB patent are unlikely.
4. Next, after lifting the mesocolon in a cephalad direction and exposing the root of the mesentery, the SMV is identified.
5. Choice of conduit: The IJV is most often used though other options have been described. These include the IMV, coronary vein, and a recanalized umbilical vein. Grafts made of synthetic material may be used in the absence of a suitable venous autograft or as an extension to a vein graft that is too short, but are not a preferred option. The IJV is removed by taking the length of the vein from the jugular foramen to the thoracic inlet.
6. The IJV to intrahepatic left portal vein anastomosis is performed first. The back wall is performed first using either 7/0 or 8/0 Prolene® running suture under loupe magnification. The anterior wall anastomosis is completed using 7/0 or 8/0 Prolene® interrupted sutures. The authors have found that this technique has helped reduce the incidence of postoperative anastomotic stricture, as it allows the graft to

expand more. The anterior aspect of the graft should be marked to help prevent further twisting of the graft as it will be tunneled.

7. The graft is tunneled through the hepatogastric ligament, passing posteriorly to the stomach, anteriorly to the pancreas, and then through the base of the transverse mesocolon until reaching the SMV. Great caution is used to avoid twisting the graft. The anastomosis is performed using 6/0 Prolene□ running suture and a "growth factor" should be used to allow expansion of the graft.
8. After releasing the vascular clamps, the flow through the graft is evaluated and quantified using vascular flow probes (Transonic□). Values obtained are compared with ideal expected portal venous flow, based on cardiac output. The calculation method of the ideal portal venous flow has been presented in a previous publication by Superina et al., 2006 (see "Further reading"). If the measured flow value differs greatly from expected flow or is less than 40% of calculated ideal portal vein flow based on the body surface are of the child, factors that may be accounting for the less than ideal flow are identified and corrected. Common problems to exclude include vein graft twisting as it is tunneled; extrinsic compression of the graft through the tunnel; anastomotic complications at the hepatic end anastomosis of the graft. Platelet thrombi may develop and a thrombectomy should be performed using Fogarty balloon catheters. Tissue plasminogen activator (tPA) may also be injected. It may be necessary to perform an intraoperative venogram to better identify the source of the problem. However, the most common factor identified that accounts for poor vein graft flow is the quality of the left portal vein in the liver. Marginal vein quality as defined by damaged endothelium or tiny variceal like branches cannot sustain forward blood flow in the graft, and inevitably it will thrombose even with full anticoagulation.

Distal splenorenal shunt

The end-to-side distal splenorenal shunt (Warren's shunt) (**Figure 53.5a and b**) is a selective portosystemic shunt, and is associated with a lower incidence of postoperative hepatic encephalopathy than the central splenorenal shunt (non-selective shunt) or the side-to-side splenorenal shunt. In contrast to the MRB, it does not restore mesenteric venous blood flow to the liver, but it effectively decompresses both gastric and esophageal varices, reducing the risks of future GI bleeds. It also decompresses the spleen, improving or eliminating splenomegaly and hypersplenism. The DSRS is the procedure of choice for the management of PHT in patients with intrinsic liver disease without evidence of advanced liver disease.



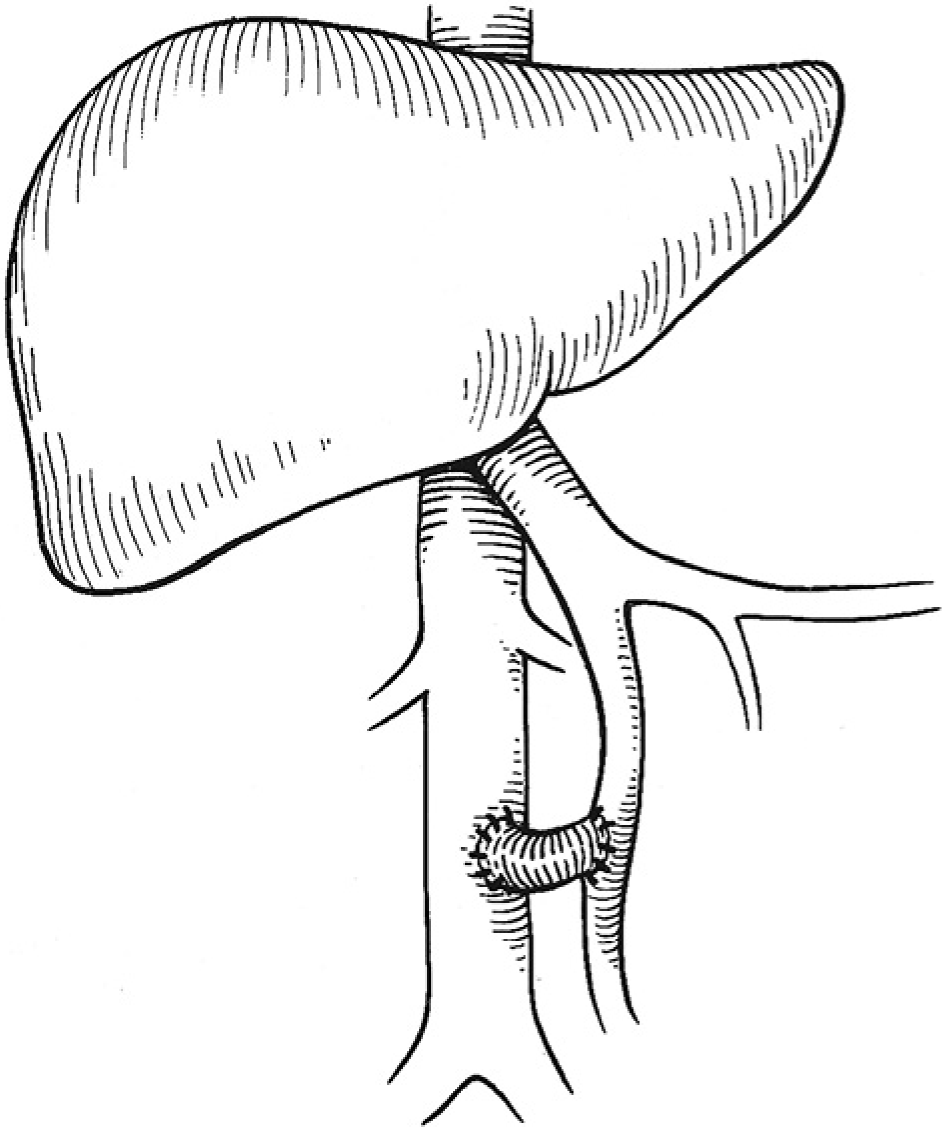
53.5a,b (a) Distal splenorenal shunt, schematic representation. (b) Distal splenorenal shunt, intraoperative picture. The suction device is lifting the pancreas, exposing the vascular anastomosis (black arrow). (SV, splenic vein; LRV, left renal vein.)

1. After accessing the intraperitoneal cavity (midline or left subcostal incision), the lesser sac is entered. The splenic vein is identified under the inferior border of the pancreas. It is dissected proximally until reaching the confluence with the SMV. The SMV end of the splenic vein is clamped, and oversewn with a 5/0 or 6/0 Prolene running suture. Distally, over-dissection of the splenic vein must be avoided as it may cause twisting or kinking of the vein. The dissection should only be carried out in order to obtain enough length for the splenic vein to reach the left renal vein without tension.

2. The left renal vein is identified also from within the lesser sac using a combination of palpation (identifying the position of the left kidney, and the pulsations of the aorta and left renal artery) and intraoperative Doppler US. The ligament of Treitz is identified, and care is taken not to go through the transverse mesocolon (possible site of internal hernias). The adrenal and gonadal veins are isolated and encircled with vessel loops. The renal vein is freed circumferentially, and dissected medially to increase its mobility.
3. The adrenal and gonadal veins are double looped, and the renal vein is clamped using a medium clamp. The splenic vein end is spatulated, and after ensuring that it is not twisted, and end-to-side anastomosis is performed with 6/0 Prolene□ running sutures. A “growth factor” should be used to allow expansion of the graft.
4. Flow through the shunt is assessed using flow probes and intraoperative Doppler US.
5. Lastly, the coronary vein is identified, ligated, and divided.

Mesocaval shunt

The MCS (**Figure 53.6**) is usually not a first option in the management of PHT in children, as it may be associated with a greater degree of postoperative hepatic encephalopathy and higher ammonia levels. However, it may be the only surgical option for some in whom neither a MRB nor DSRS can be performed (e.g. cases of portal vein thrombosis extending in the splenic vein, or patients who have undergone a splenectomy). In the absence of intrinsic liver disease, a non-selective shunt rarely causes aggravation of encephalopathy, as it may in those with advanced liver disease, and may be performed safely. A word of caution is necessary: In children whose symptoms include portopulmonary hypertension or hepatopulmonary syndrome as a consequence of shunting, PSS is contraindicated and should not be done to treat symptomatic PHT.

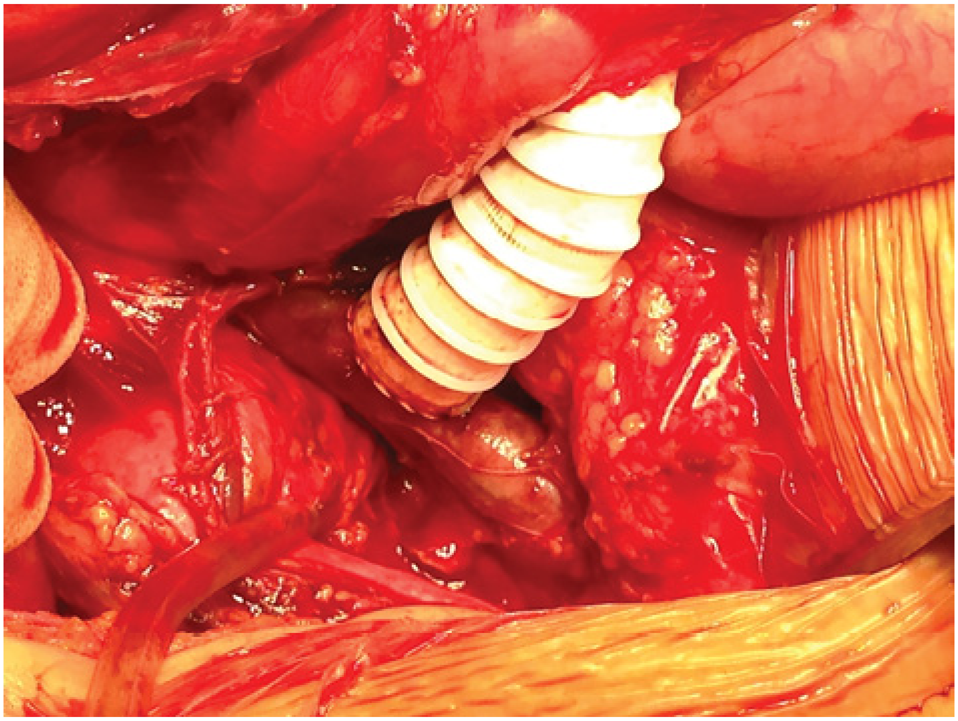


53.6 MCS, schematic representation.

1. The abdominal cavity is entered through a midline incision. The transverse colon is reflected cephalad. The root of the mesentery is palpated to identify the pulsation of the superior mesenteric artery (SMA). The SMV is located to its right. Intraoperative Doppler US can also be used to identify the SMV. The vein is exposed, and its main branches are encircled with vessel loops.
2. A window is made through the bowel mesentery, through which the IVC is identified and isolated as it emerges from behind the duodenum.

Sufficient length must be dissected in order to safely apply the vascular clamp required for the confection of the anastomosis.

3. Choice of conduit: The authors favor the use of a reinforced Gore-Tex® graft to perform an MCS (**Figure 53.7**). Venous autografts such as the IJV are more compressible, and are at increased risk of kinking leading to shunt obstruction as they traverse the bowel mesentery between the SMV and IVC once the bowel is placed back in the abdominal cavity.



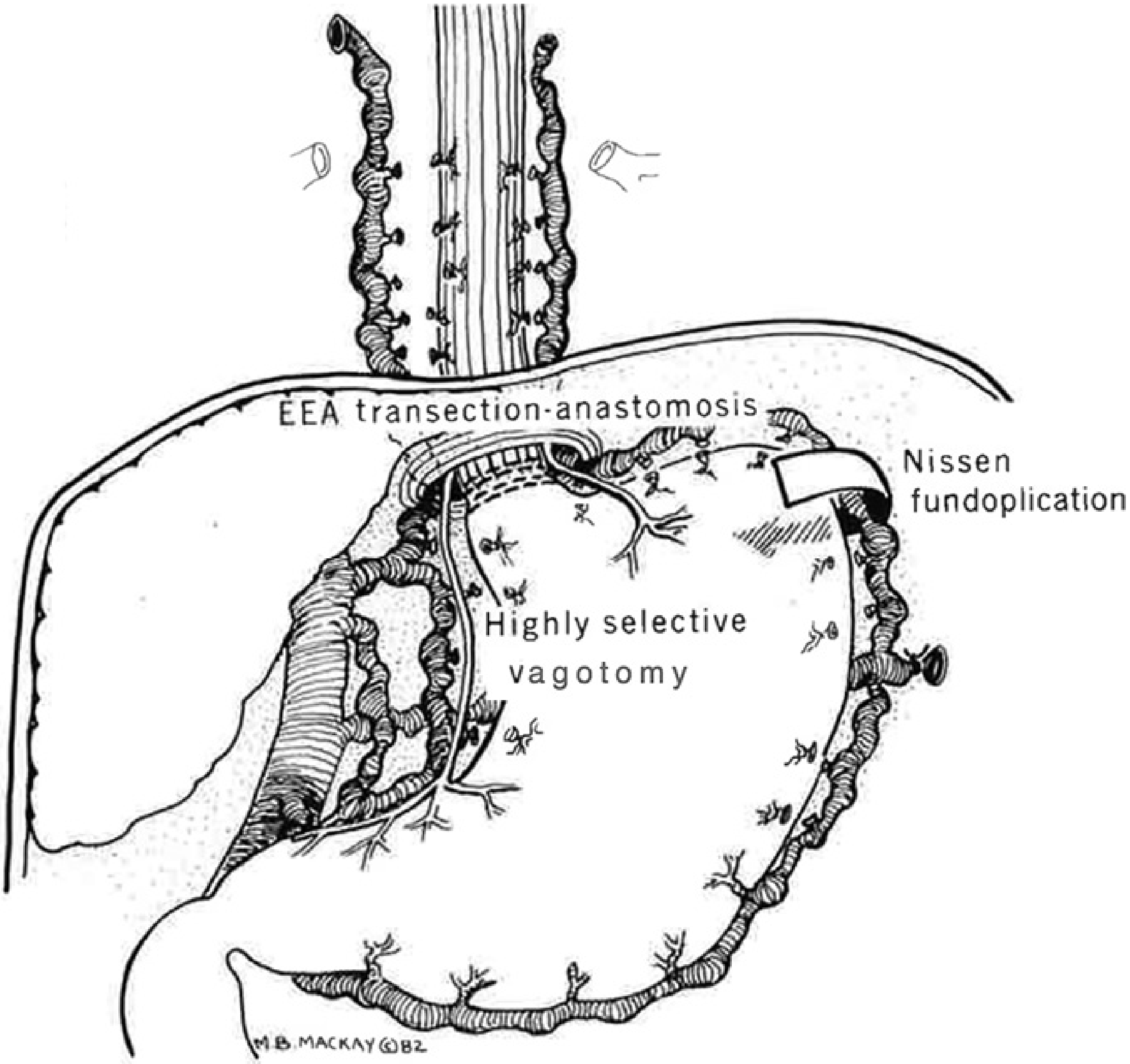
53.7 Intraoperative picture of an MCS; Gore-Tex® graft.

4. The vena cava is clamped with a medium vascular clamp, and a venotomy is performed. An end-to-side anastomosis is performed first between the graft and the IVC using 5/0 Prolene® running sutures.
5. The SMV is then controlled using a fine vascular clamp. The Gore-Tex® graft is trimmed so it reaches the SMV without tension or redundancy. It is anastomosed to the SMV using 6/0 Prolene running sutures. The vascular clamps are released. Unfortunately, the use of synthetic material prevents the use of vascular flow probes to assess flow through the shunt.

6. Lastly, the coronary vein is identified and ligated.

Sugiura procedure

The esophagogastric devascularization procedure (Sugiura procedure) (**Figure 53.8**) is usually indicated to effectively control variceal bleeding when there is no other surgical portosystemic shunt option possible (extensive intra-abdominal venous thrombosis, for example in cases of hypercoagulable conditions). The authors favor a modified Sugiura procedure, in which the lower esophagus is transected with a circular stapler, and the spleen is preserved.


EEA transection-anastomosis
Nissen
fundoplication
Highly selective
vagotomy
M.B. MACKAY ©82

53.8 Modified Sugiura procedure.

1. The procedure can often be performed successfully through a midline abdominal approach alone. A thoracoabdominal incision ensures optimal visualization of the proximal periesophageal variceal network in the chest, but it is often not necessary in the pediatric population, particularly small children.
2. Using a combination of a vessel-sealing device e.g. LigaSure (Valleylab, Boulder, CO, USA), Harmonic Scalpel (Ethicon Inc., Cincinnati, OH, USA) and tie ligation, the large varices along the greater and the lesser

curvature of the stomach are divided until reaching the esophagogastric junction. The short gastric vessels to the spleen are also therefore divided. The dissection continues above the junction into the chest, as far up as is possible, optimally until reaching the inferior pulmonary vein. The innervation to the pylorus and the antrum is preserved.

3. A gastrotomy is made, through which a circular stapling device is introduced and applied to the esophagus above the gastroesophageal junction. This maneuver interrupts the intraesophageal mural varices. A fundoplication is then easily performed over the stapled suture line, as the short gastric vessels have been divided from the fundus. It effectively protects the staple line if there is any concern with a possible leak. A Heineke–Mikulicz pyloroplasty is also performed because of the possible occurrence of delayed gastric emptying.

POSTOPERATIVE CARE

Patients undergoing an MRB or portosystemic shunt procedure are placed on a prophylactic heparin infusion with the aim of not increasing the partial thromboplastin time (PTT). If the patient has a known hypercoagulable condition of if the surgeon believes that the MRB is more susceptible to thrombosis than usual, a therapeutic heparin infusion can be started with the PTT goal of 60–90 seconds, or 2–2.5 times the normal laboratory value. A Doppler US is performed on the first postoperative day. If vascular graft or shunt patency is confirmed, the patient is started on an oral Clopidogrel 1 mg/kg once a day and allowed a clear fluid diet. An abdominal CT scan with IV contrast (portal venous phase) is performed 7 days after the surgery.

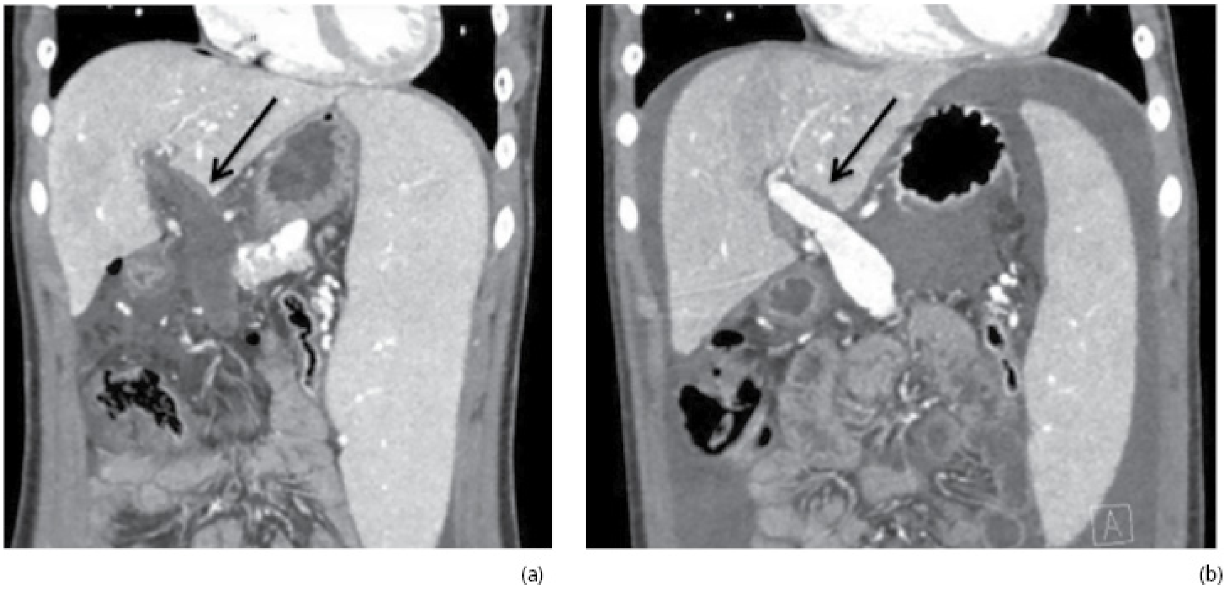
Uncomplicated patients are followed as outpatients 1 month after surgery, then every 3 months for a year, and then yearly for 5 years. Resolution of splenomegaly is monitored through serial physical examination. A CBC, an ammonia level, and an abdominal Doppler US are performed at each appointment. The US evaluates both the patency of the shunt, and the velocities through the anastomoses. If there are no complications, a magnetic resonance imaging study is performed at the 1-

year mark. This study provides an estimated blood flow through the venous conduit. Antiplatelet therapy is continued for 6 months after the surgery and then usually discontinued. Patients who suffer postoperative complications or have a known underlying hypercoagulable condition may require more frequent follow-up and cross-sectional imaging, and prolonged anticoagulation and/or antiplatelet therapy.

COMPLICATIONS

Thrombosis

Thrombosis can occur during or soon after the surgery (**Figure 53.9a and b**). The anastomosis may need to be revised. Fogarty catheters can be used to perform a thrombectomy, and tPA may need to be injected. Those patients who suffer from a thrombotic event are fully anticoagulated (oral warfarin (Coumadin□) or subcutaneous enoxaparin (Lovenox□)) postoperatively. The anticoagulation is continued for at least 6 months, after which an antiplatelet agent is started and may be continued indefinitely. Late shunt thrombosis now occurs rarely with the improvement in preoperative screening for hypercoagulable conditions, but it may still occur in high-risk patients. They may present with recurrent GI bleed.



53.9a,b (a) On the first postoperative day after an MRB, the US failed to show any flow through the graft. The patient underwent a CT, showing the thrombosed shunt (black arrow). (b) Same patient, 4 days after surgical revision of the shunt, contrast can be seen inside the shunt (black arrow).

Stenosis

Stenosis tends to present within the first few months of surgery but rarely after a year. Clinical findings of a significant stenosis include persistence or recurrence of splenomegaly. A low platelet count that fails to improve or starts decreasing after initial improvement is another suggestive finding. A rising ammonia level indicates recurrence of systemic venous shunting. US may also be suggestive of stenosis with increased blood velocity and turbulence through an anastomosis. A CT scan is obtained to confirm the diagnosis. Interventional radiology anastomotic angioplasty is the treatment of choice for this condition. The patient is placed on antiplatelet therapy following the procedure. A recurrent stricture despite angioplasty may require stenting or surgical revision. Stenting is avoided in younger patients for recurrent strictures until a stent may be placed that will serve the child into adulthood and not represent a fixed stenosis that will be unable to be dilated further as the child grows over time.

OUTCOME

We have performed over 200 MRB operations for idiopathic portal vein thrombosis in children. Approximately 70–80% of all children with EHPVO referred to our institution for an MRB will ultimately undergo a successful MRB. The other 20–30% of the children who do not qualify for this operation do so principally because of damage to the portal vein that extends into the liver itself and an intrahepatic anastomosis is not possible. These patients for the most part can be treated with a DSRS or an MCS. In a review of our institutional experience with patients with EHPVT in 2010, we reported that both the MRB and other portosystemic shunts were effective at curing GI bleeding in pediatric patients with EHPVO. However, the MRB was superior at relieving hypersplenism. By restoring normal blood flow to the liver, the MRB was also found to have positive metabolic effects. Growth and neurocognitive abilities were both improved post-procedure. Unpublished data also suggest that MRB in patients with pulmonary hypertension may help stabilize pulmonary hypertension.

Patients with PHT from stable parenchymal liver disease who receive DSRS also experience long-term patency of the shunt, stabilization and improvement of hypersplenism, and cessation of bleeding from esophageal and gastric varices or stable parenchymal disease.

TIPS is rarely carried out in our institution other than as a palliative measure in preparation for a transplant.

FURTHER READING

Di Francesco F, Grimaldi C, de Ville de Goyet J. Meso-Rex bypass – a procedure to cure prehepatic portal hypertension: The insight and the inside. *J Am Coll Surg* 2014; 218(2): e23–36.

Lautz TB, Keys LA, Melvin JC, et al. Advantages of the meso-Rex bypass compared with portosystemic shunts in the management of extrahepatic portal vein obstruction in children. *J Am Coll Surg* 2013; 216(1): 83–9.

Lautz TB, Kim ST, Donaldson JS, Superina RA. Outcomes of percutaneous interventions for management stenosis after meso-Rex bypass for extra hepatic portal vein obstruction. *J Vasc Interv Radiol* 2012; 23(3): 377–83.

Lautz TB, Sundaram SS, Whitington PF, et al. Growth impairment in children with extrahepatic portal vein obstruction is improved by mesenterico-left portal vein bypass. *J Pediatr Surg* 2009; 44(11): 2067–70.

Ling SC. Advances in the evaluation and management of children with portal hypertension. *Semin Liver Dis* 2012; 32(4): 288–97.

Mack CL, Zelko FA, Lokar J, et al. Surgically restoring portal blood flow to the liver in children with primary extrahepatic portal vein thrombosis improve fluid neurocognitive ability. *Pediatrics* 2006; 117(3): e405–12.

Shneider BL, de Ville de Goyet J, Leung DH, et al. Primary prophylaxis of variceal bleeding in children and the role of Meso-Rex Bypass: Summary of the Baveno VI Pediatric Satellite Symposium. *Hepatology* 2016; 63(4): 1368–80.

Superina R. The meso-Rex shunt. In: Davenport M, Heaton ND, Superina R (eds). *Surgery of the Liver, Bile Ducts and Pancreas in Children*, 3rd ed. Boca Raton, FL: CRC Press; 2017: pp. 292–307.

Superina R, Bambini D, Lokar J, et al. Correction of extrahepatic portal vein thrombosis by the mesenteric to left portal vein bypass. *Ann Surg* 2006; 243(4): 515–21.

Superina RA, Weber JL, Shandling B. A modified Sugiura operation for bleeding varices in children. *J Pediatr Surg* 1983; 18(6): 764–9.

54

Cholecystectomy

MARK DAVENPORT

HISTORY

Gallstones have been known about since time immemorial with examples being found in Egyptian mummies. The first successful cholecystostomy was performed in 1867 when John Stough Bobbs from Indianapolis removed a gallstone in a 30-year-old woman with cholecystitis. Definitive open cholecystectomy was first performed by the German surgeon Carl Langenbuch in July 1882.

The first laparoscopic cholecystectomy was performed by another German surgeon, Erich Mühe, in September 1985, although this was not reported at the time. The report to the German Surgical Society in 1986 was shouted down and not at all well received. Little did they know they had just witnessed the future!

PRINCIPLES AND JUSTIFICATION

The main indication for cholecystectomy is the presence of symptomatic gallstones. These differ from those found in adults, which are largely mixed and cholesterol-based. In children and adolescents, over half will be black pigment stones derived from a hemolytic process, and only 25% will be cholesterol stones. Other types seen in children include calcium carbonate, protein-dominant and brown pigment stones. In many Western countries, there has also been a dramatic rise in the prevalence of typical cholesterol stones in overweight female adolescents. Table 54.1 lists some underlying causes of gallstones.

Table 54.1 **Gallstones in children**

Hemolytic	Inspissated bile/sludge	Cholesterol metabolism
Sickle cell anemia Hereditary spherocytosis (elliptocytosis) Thalassemia Gilbert's syndrome	Parenteral nutrition Ceftriaxone Rapid weight loss	Obesity Progressive familial intrahepatic cholestasis

Gallstones may cause biliary colic and acute and chronic cholecystitis while in the gallbladder; if they traverse the cystic duct and enter the common duct, they may cause obstructive jaundice, cholangitis, and, if impacting at the ampulla, acute pancreatitis.

It is possible to have gallbladder disease without gallstones though this should only be considered after appropriate investigations. Two situations are worthy of note.

- *Biliary dyskinesia*, though controversial, is being increasingly recognized as a cause of epigastric or right upper quadrant abdominal pain, nausea, and/or vomiting. If a radioisotope gallbladder-emptying scan can show an abnormal ejection fraction of <35% (or better still <10%), then cholecystectomy may be considered. Still, up to 40% of those patients will not benefit from surgery.
- *Acalculous cholecystitis* in children arises in association with and is caused by severe sepsis, trauma, or burns. Ultrasound may show evidence of the inflammatory process, with mural edema and thickening, gallbladder distention, and occasionally sludge. It should be treated conservatively in the first instance though acute decompression with a cholecystostomy may be considered.

The diagnostic workup should always include ultrasonography, liver biochemistry (especially conjugated bilirubin and γ-glutamyl-transpeptidase), and a search for underlying metabolic, liver disease, or structural malformation of the biliary tree. It is also important to identify or exclude choledocholithiasis prior to surgery. Detailed ultrasonic imaging of the common hepatic and bile ducts should detect a dilated bile duct with intraluminal stones or debris. Where doubt exists, a magnetic resonance cholangiopancreatography (MRCP) should provide adequate anatomical detail.

PREOPERATIVE PREPARATION

Most children will not require specific special preparation. However, the exceptions to this are children with sickle cell disease (SCD) as they are at great risk of acute vaso-occlusive pain crises, acute chest syndrome, and seizures developing postoperatively. There may also be problems with cross-matching because of a history of multiple blood transfusions and alloimmunization together with relative immunosuppression due to asplenia arising as result of chronic splenic infarction. Table 54.2 illustrates a suggested perioperative preparation for a child with SCD.

Table 54.2 **Perioperative care of the child with SCD**

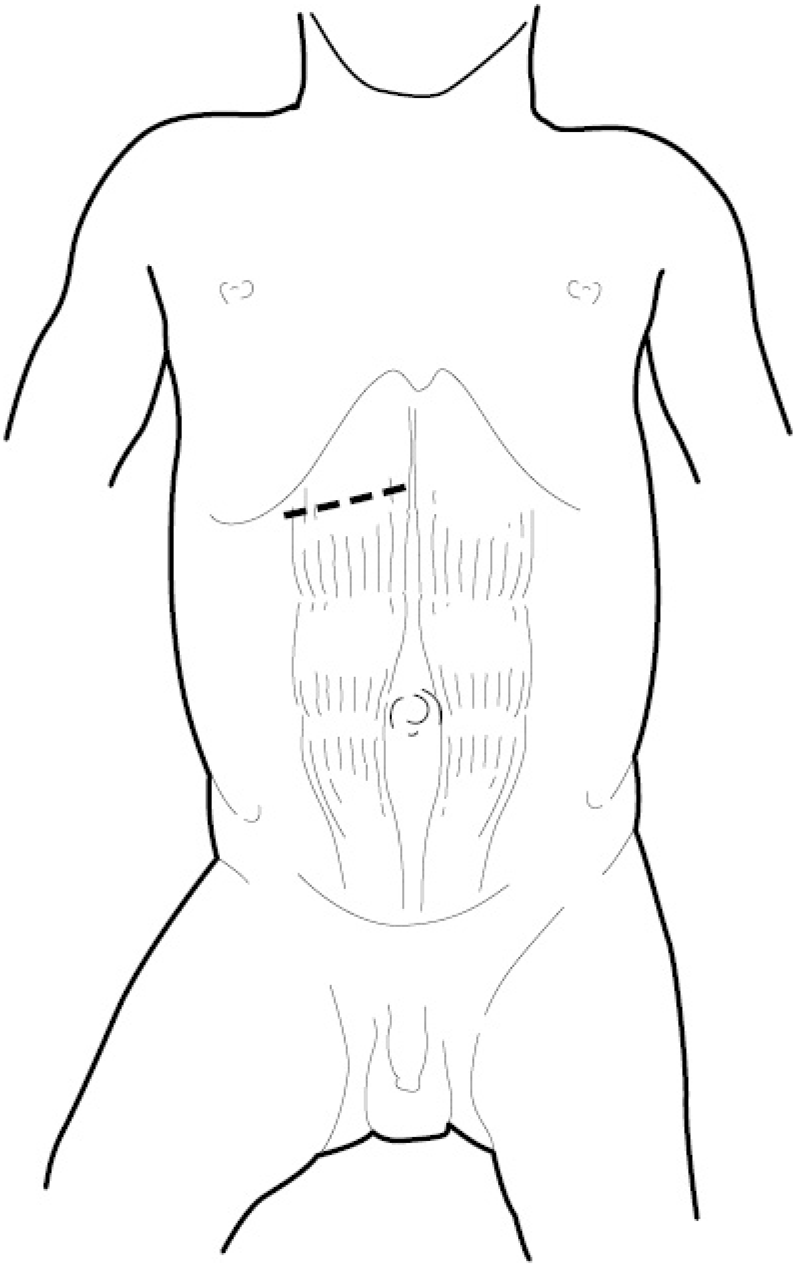
Preoperative	
Transfusion (simple as good as exchange)	Aim to achieve ≥100 g/L
Intravenous rehydration	12–24 hours pre-anesthesia – NaCl (0.9%) IV
Operation and anesthesia	
Pre-oxygenation	Nasal cannulas
Maintenance of body temperature	Body-warming devices, heated IV fluids
Maintenance of peripheral warming	Pressure area care
Avoidance of tourniquets	
Postoperative	
Maintain O_2 saturation	Supplemental O_2
Chest physiotherapy	Avoid basal atelectasis to minimize risk of acute chest syndrome
Effective analgesia	To encourage coughing and clearance of secretions Possible opiate tolerance due to preoperative use for "crises"
Early mobilization	

OPEN CHOLECYSTECTOMY

Though this is less commonly performed than the laparoscopic alternative, there may be circumstances, such as lack of laparoscopic experience or appropriately sized equipment, in which this will be the better option. Additionally, in those with extensive previous surgery and dense adhesions, the open option may give a safer dissection. Prophylactic antibiotics should be given.

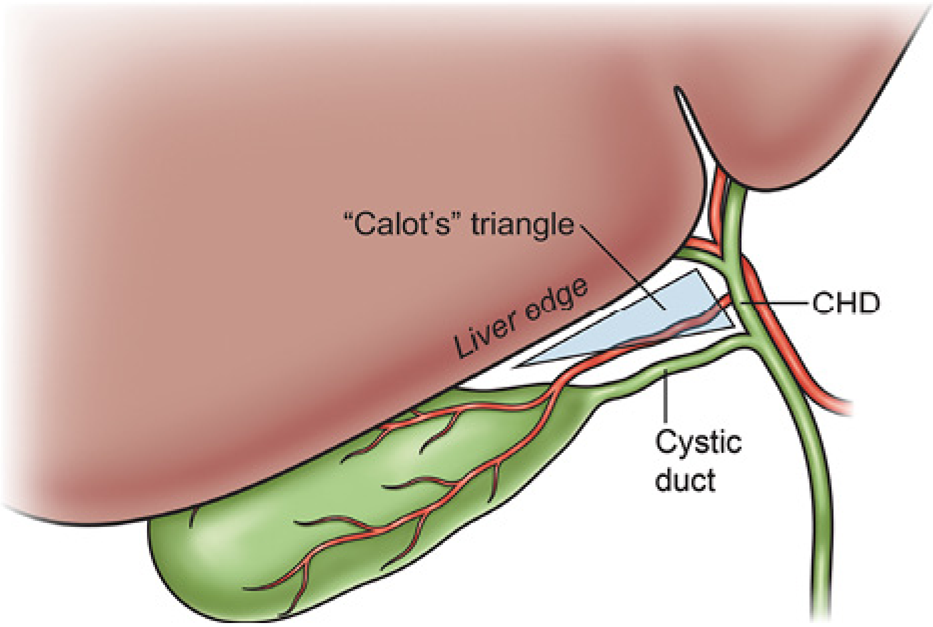
Key operative steps

A right upper quadrant muscle-cutting incision is made (**Figure 54.1**). Two abdominal gauze packs are inserted over the stomach and duodenum, and the transverse colon, held by the left hand of the assistant to expose the gallbladder, its arterial supply, and junction of cystic duct with common bile duct (CBD). The fundus of the gallbladder is retracted laterally with a Babcock□ forceps or by traction on a stay suture. Cephalic retraction of the liver can be achieved by use of malleable copper or Deaver-pattern□ retractors.

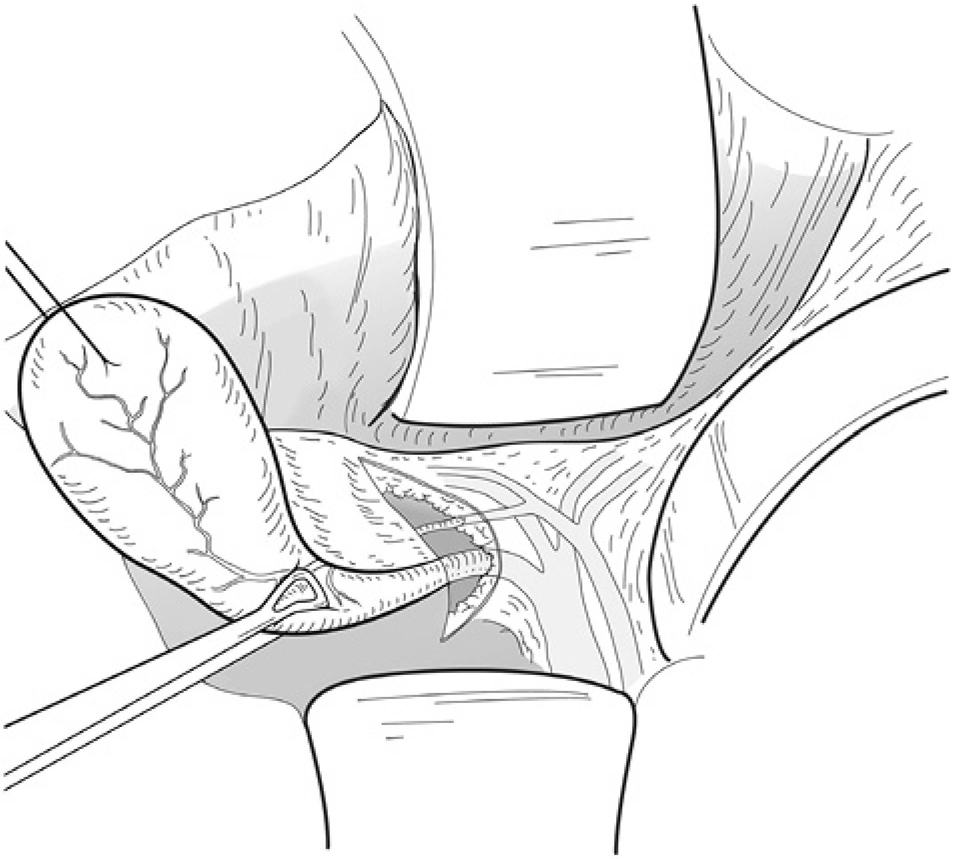


54.1

Traditionally, the dissection starts in Calot's triangle, named after French surgeon Jean-François Calot (1861–1944), to identify the cystic artery and cystic duct with orthograde removal of the gallbladder (**Figure 54.2**). This is best achieved by use of a non-toothed "vascular" tissue (e.g. DeBakey™) and right-angled dissecting forceps (e.g. Lahey™, Mixter™). In older children, lateral traction can be achieved by Babcock□ forceps on Hartmann’s pouch (**Figure 54.3**).


“Calot’s” triangle
Liver edge
CHD
Cystic duct

54.2 “Calot’s” triangle: Cystic duct, common hepatic duct (CHD), and edge of liver.

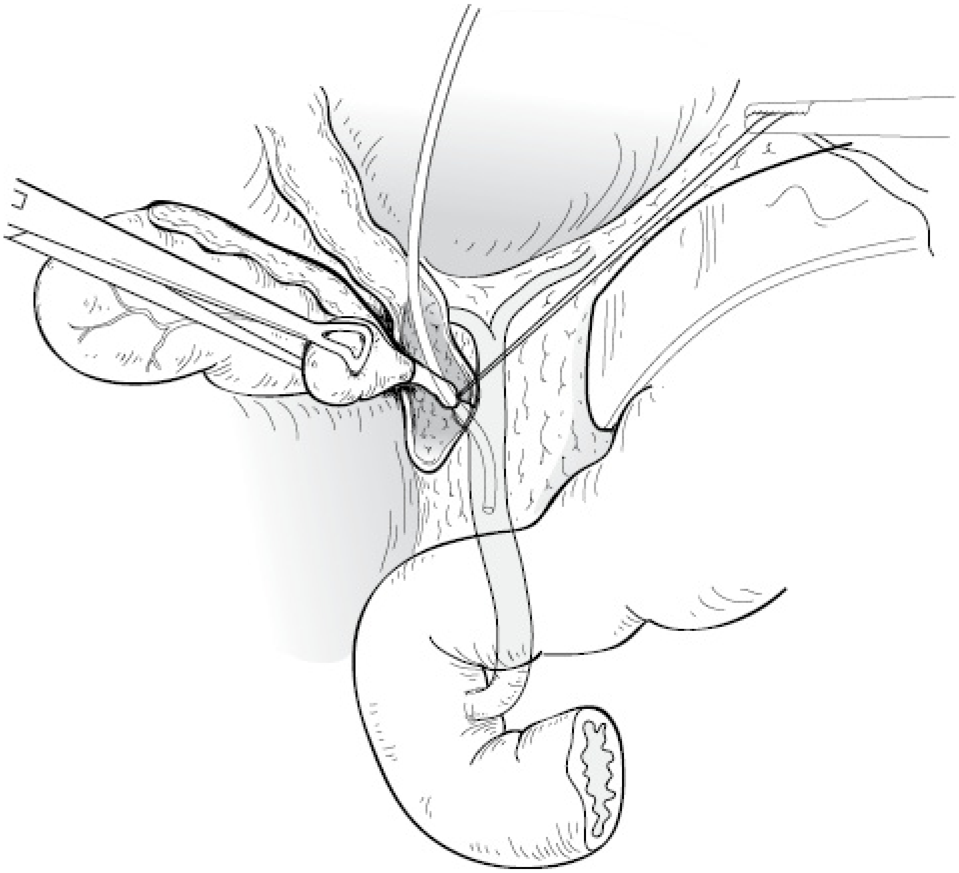


54.3

The aim (in both open and laparoscopic operations) is to show the contents of this triangle clearly and to establish a “critical view of safety” before committing to any actual division of key structures. This concept was introduced in 1995 after the first wave of laparoscopic cholecystectomies in the early 1990s was shown to be associated with a higher incidence of bile duct injury. There are three key elements:

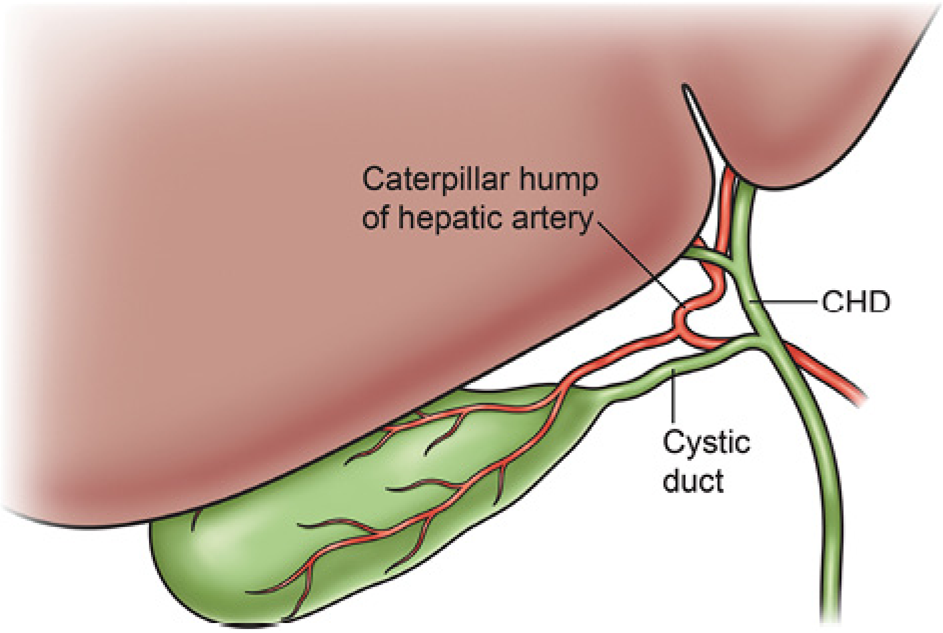
1. Calot’s triangle is cleared of fat and fibrous tissue. Neither CBD nor CHD have to be exposed.
2. The lower third of the gallbladder is separated from the liver (anterior and posterior) to create a window.
3. This should leave only two structures entering the gallbladder (and nowhere else).

An on-table cholangiogram is relatively easy to perform during open surgery compared to the laparoscopic approach. A small incision is made in the proximal cystic duct. Insert an appropriate cholangiogram catheter (e.g. Stoke-on-Trent™) or, more usually because of size constraints, an “umbilical vein” catheter (**Figure 54.4**). Both should be prefilled with saline and bubble-free. Any difficulties with insertion can usually be overcome with probing (e.g. “lacrimal duct” probes). Once the catheter is correctly sited, the contrast solution can be injected. The cholangiogram should show left and right hepatic ducts, the intrahepatic ducts, and free-flowing exit into the duodenum. Any filling defects can be presumed to be stones.



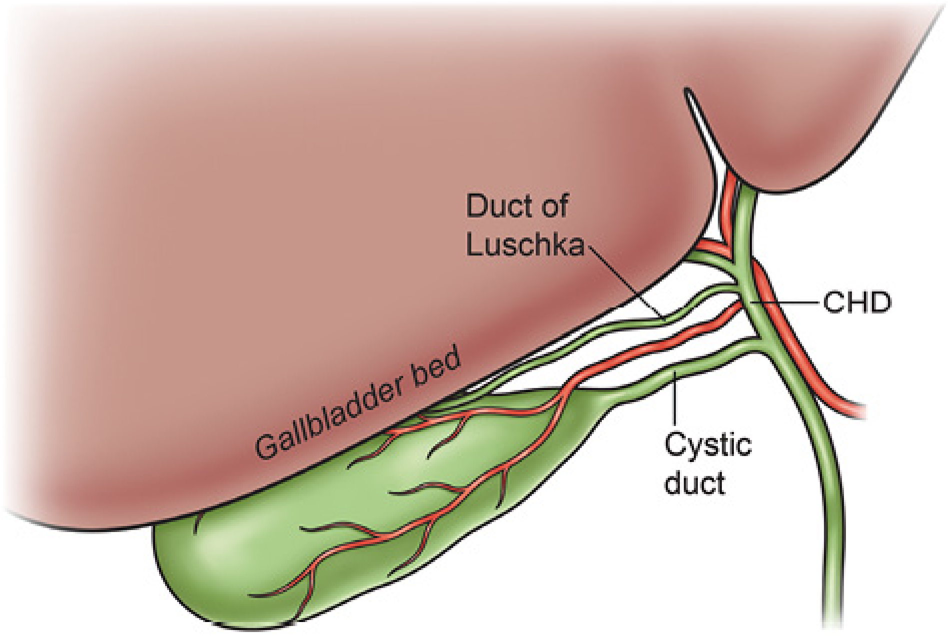
54.4

The cystic artery usually traverses Calot’s triangle arising from an anterior right hepatic artery. It can be ligated and divided safely only if it can be shown to be supplying the gallbladder (and nothing else). There are many variations of arterial anatomy which can confuse and render the right hepatic artery at risk. One such is the caterpillar hump (of Moynihan) configuration, making it unduly prominent in Calot's triangle (**Figure 54.5**). Dissection of the cystic duct down to the junction with CBD is relatively straightforward being in continuity with Hartmann's pouch. Nonetheless, variations include a low insertion near the duodenum or even within the pancreas. Where there is doubt, a cholangiogram is indicated. Both cystic duct and artery can be divided and ligated with absorbable material.



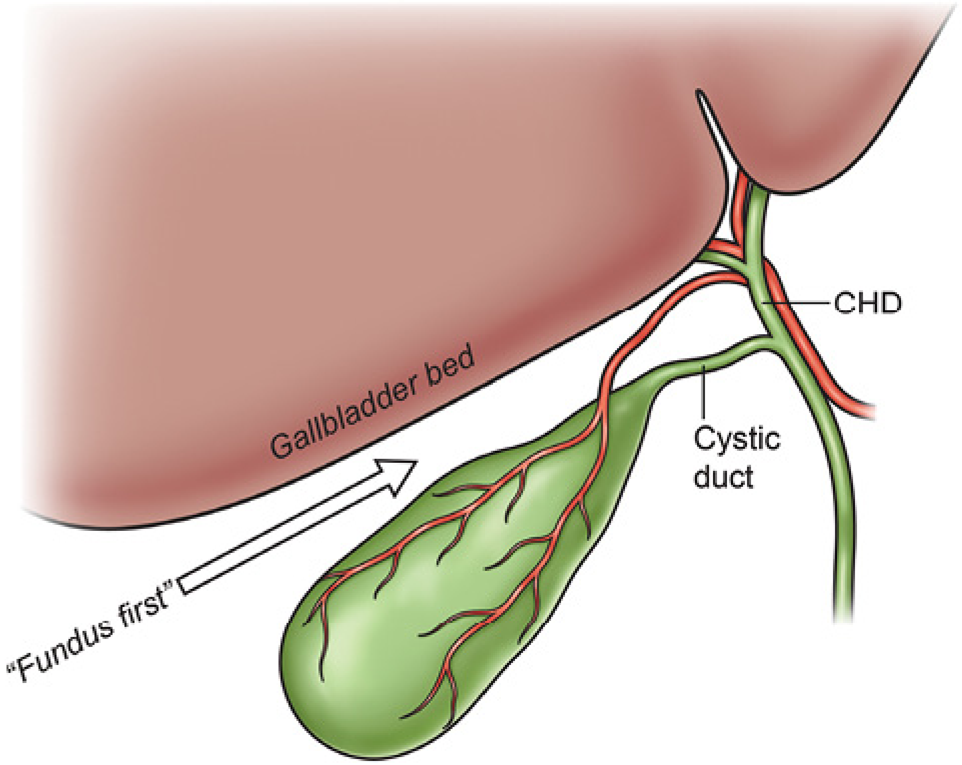
54.5 "Caterpillar Hump" of right hepatic artery.

Hartmann's pouch should now be completely free and the gallbladder can be completely dissected off its bed, typically using diathermy to control any bleeding from the multiplicity of veins. Sometimes, a small bile leak can be appreciated from the bed due to division of a cholecystohepatic duct or damage to a duct of Luschka (**Figure 54.6**). This should be oversewn. Drains may be left according to preference but are not mandatory.



54.6

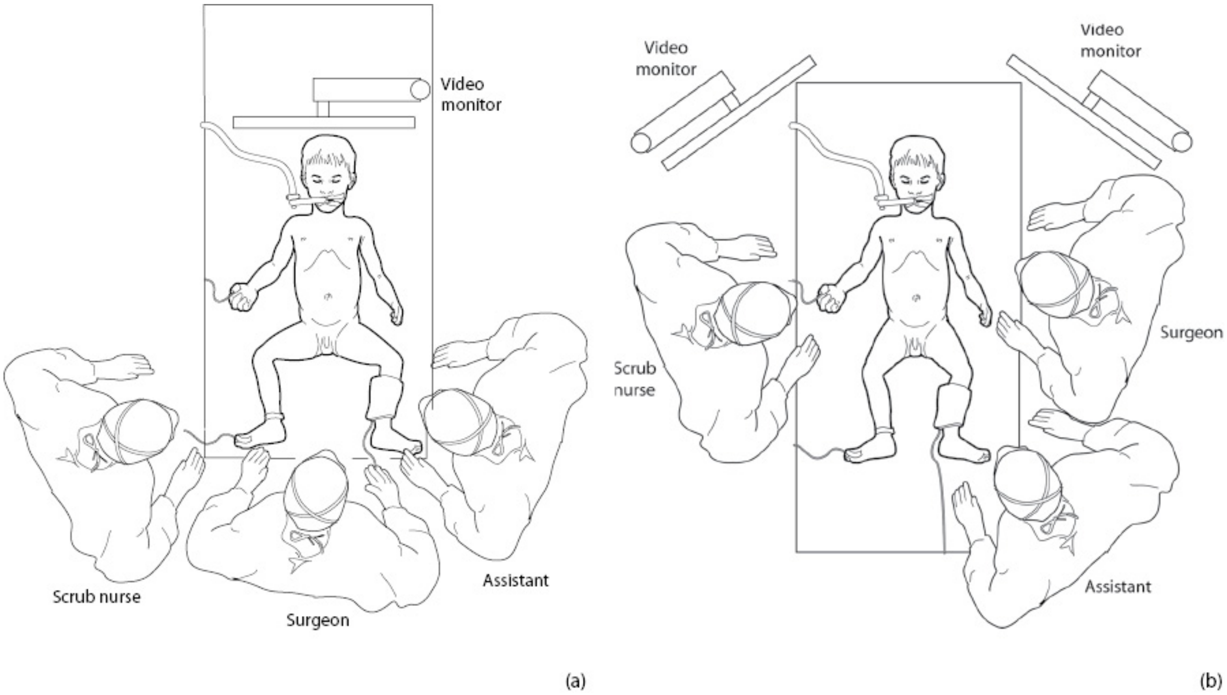
In some cases, initial Calot dissection may be challenging and not at all safe because of dense adhesions. In such cases, retrograde (“fundus-first”) cholecystectomy can be safer (**Figure 54.7**). The key is to keep the dissection on the wall of the gallbladder. Calot’s triangle is opened from above and the cystic artery should be the first structure encountered. Eventually, the gallbladder should narrow until the cystic duct is the only thing left.


Gallbladder bed
CHD
Cystic duct
"Fundus first"

54.7 "Fundus-First" approach.

LAPAROSCOPIC CHOLECYSTECTOMY

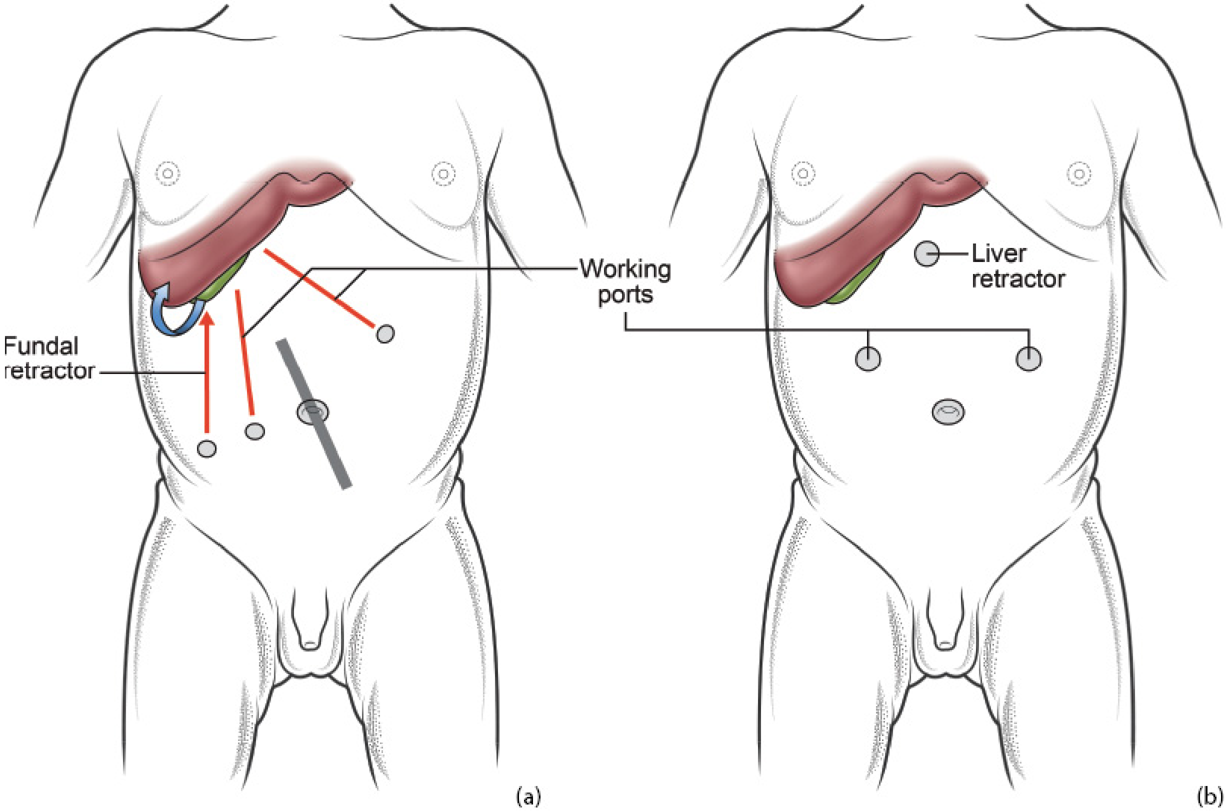
A bipolar approach to surgeons' position developed in the late 1980s and early 1990s fairly independently on either side of the Atlantic. Thus, the surgeon stands between splayed legs of the patient in the "French" approach (**Figure 54.8a**), while the surgeon stands on the patient's left in the "American" approach (**Figure 54.8b**).


Video
monitor
Scrub nurse
Surgeon
Assistant
(a)
Video
monitor
Video
monitor
Scrub
nurse
Surgeon
Assistant
(b)

54.8a,b (a) The "French" approach. (b) The "American" approach.

Port placement

Typically, three working ports are used with an umbilical camera port (5 mm or 10 mm). Our preference is for a left-sided upper quadrant for the active dissecting tool (usually the diathermy hook) with two right lower quadrant ports for fundus and Hartmann's retraction (**Figure 54.9a**). Others favor an epigastric port for elevation and retraction of the liver and two para-median ports for dissection and gallbladder retraction (**Figure 54.9b**).

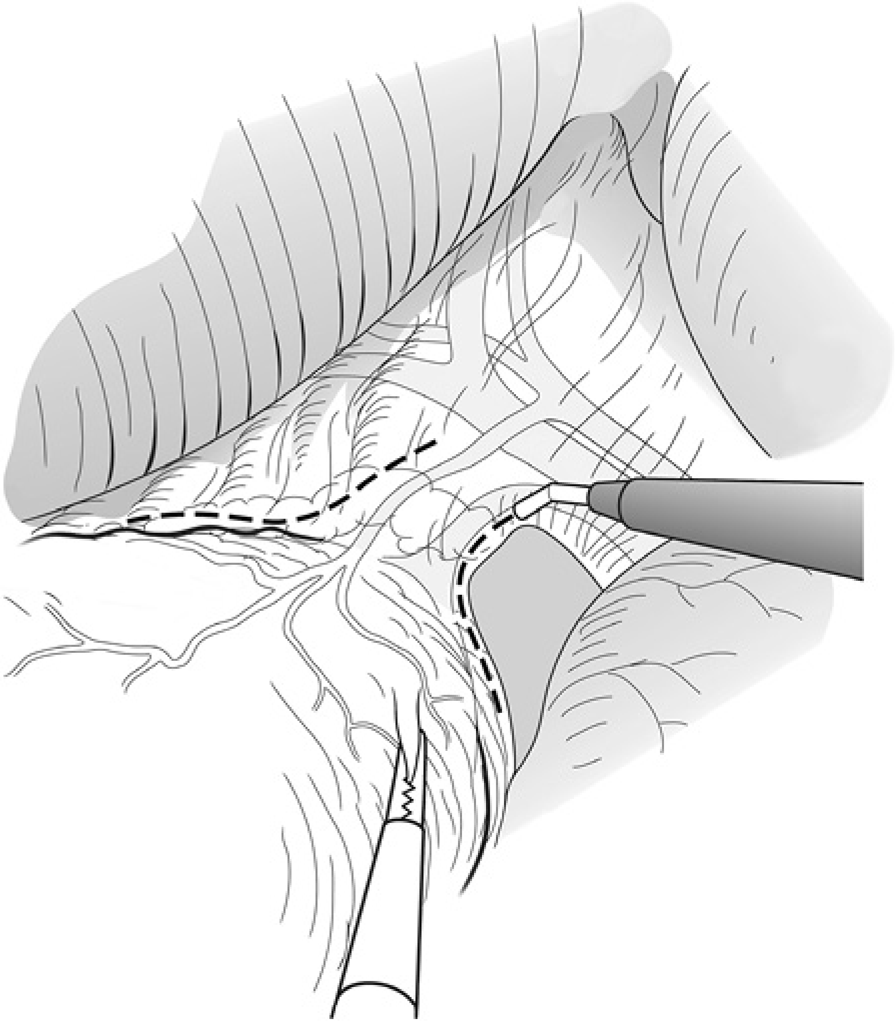


54.9a,b

A single-incision approach has been used in adolescents as it has in adults but this really compromises manual dexterity and traction for what are barely perceptible scars from the auxiliary ports.

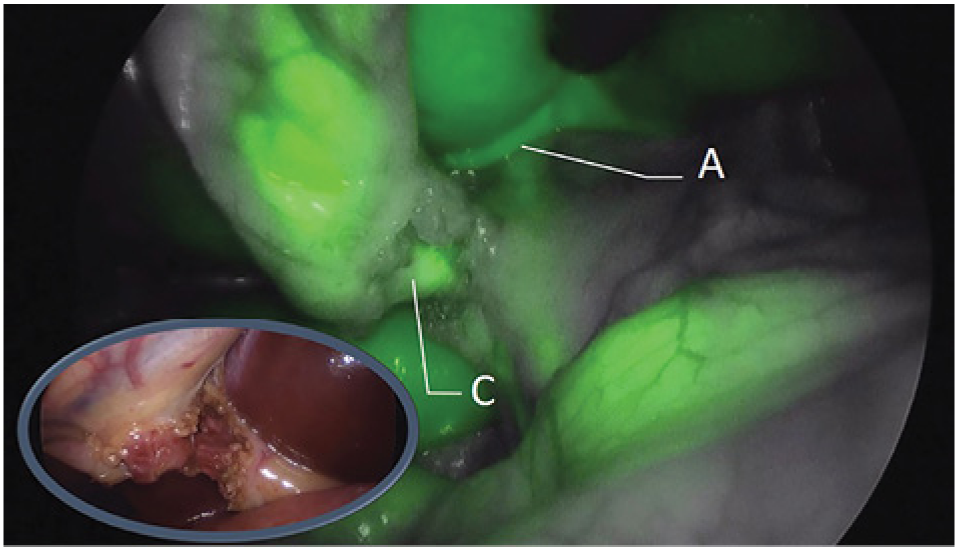
Exposure of Calot's triangle

Exert downward traction on Hartmann's pouch with "bowel grasping" forceps and craniolateral retraction on the fundus with "crocodile" forceps (**Figure 54.10**). This opens Calot's triangle and allows initial division of the overlying peritoneum using a monopolar hook. A similar dissection on the reverse aspect can be achieved by flipping Hartmann's pouch upward. The aim is to create a window between the gallbladder and liver first, displaying the cystic artery and duct. Confirm that these structures have nowhere else to go but the gallbladder and dissect around to isolate individually using dissecting (e.g. Maryland) or right-angled forceps.



54.10

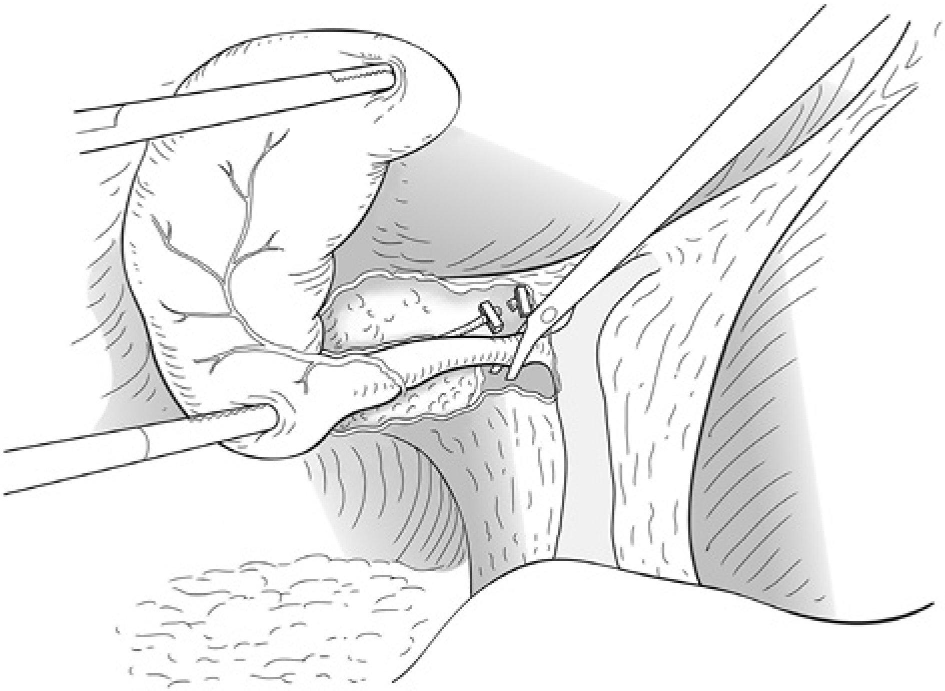
If required, a cholangiogram can be done using modified grasping forceps for Hartmann's pouch (e.g. Olsen-Reddick clamp) inserted into the lateral port after milking any stones back into the gallbladder and applying a proximal LIGACLIP™. The use of IV instillation of indocyanine green (ICG) is currently being investigated as a substitute for invasive cholangiograms. This "tags" the bile-containing structures and fluoresces under ultraviolet light so showing in great detail the CHD, CBD, and relationship to the dissection (**Figure 54.11**).



54.11 Contrast uptake can be observed initially in the cystic artery (A) and later in the cystic duct (C). (Reproduced from Fernández-Bautista B et al., *European J Pediatr Surg Rep* 2019; 7: e43–e46. doi: 10.1055/s-0039-1692191. With permission.)

Division of cystic artery and duct

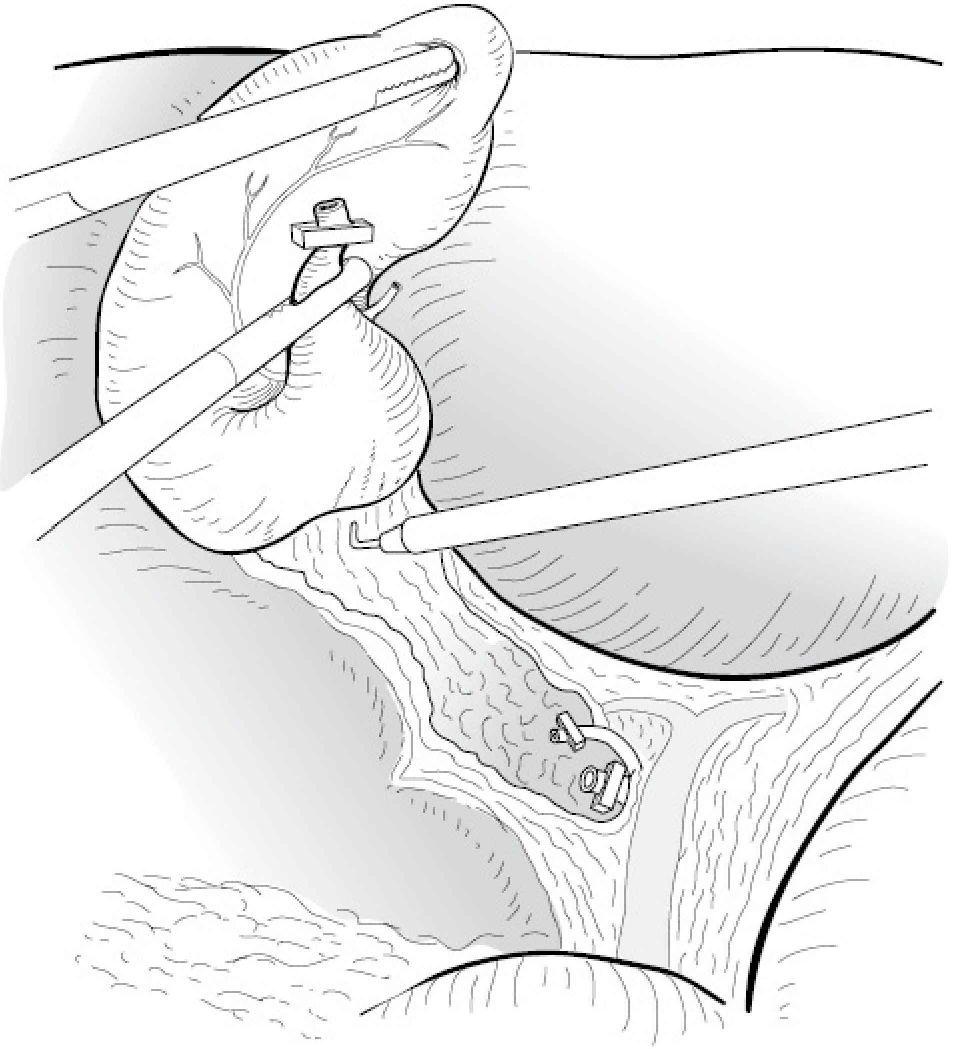
Typically, the artery is clipped (e.g. multiple LIGACLIP®) and divided leaving only the cystic duct (**Figure 54.12**). This is secured again with multiple clips or a Hem-o-lok™ and divided. Actual demonstration of the junction with CBD is not necessary but security of closure is.



54.12

Gallbadder dissection

The gallbladder can then be dissected off the bed using diathermy to control bleeding (**Figure 54.13**). Just prior to detachment, confirm that the formerly clipped artery and duct are secure. The organ can be extracted via the umbilical port by switching the camera and using a closable bag to minimize bile leakage.



54.13

Problems are usually due to inadequate dissection and visualization of key structures. A hydropic gallbladder may need to be aspirated percutaneously under vision to allow grasping forceps to take hold. If the contents of Calot's triangle are still obscure, further division of the peritoneum at gallbladder–liver interface anteriorly and posteriorly toward the fundus should create a safer zone of dissection away from the porta. Sometimes the cystic duct is too wide for safe clipped closure and may have to be ligated using a Vicryl ENDOLOOP® or even stapled across.

Choledocholithiasis

This is a presenting feature in about 10–20% of children. Our preference is to perform preliminary endoscopic retrograde cholangiopancreatography (ERCP) and duct clearance (± sphincterotomy or stenting) with a laparoscopic cholecystectomy 2–3 weeks later. Actual laparoscopic therapeutic duct exploration is rarely performed in children because the duct is not large enough to enable safe instrumentation. Nevertheless, it has been described using Dormia-basket stone retrieval and even custom-made cholangioscopes inserted through a dilated cystic duct.

COMPLICATIONS

Bile leak

These should be suspected in children after cholecystectomy with untoward abdominal pain ± jaundice. An ultrasound (US) should define a fluid collection and if there is doubt a radio-isotope scan (e.g. HIDA) can be definitive. There are three possibilities:

- Slipped clips from cystic duct
- Major bile duct injury (<2% overall)
- Inadvertent injury to congenital cholecystohepatic ducts or duct of Luschka

In our practice early anatomical definition using ERCP (± stenting) will suggest an appropriate management plan. This may simply involve continuation of stenting in the case of slipped clips but a rescue hepaticojejunostomy-en Roux may also be warranted for major common hepatic or CBD injury.

VIDEOS

OPS_8e_PPT_54.1 Cholecystectomy
OPS_8e_video_54.2 Cholecystectomy

FURTHER READING

Bean JF, Barsness KA. Cholecystectomy and surgical exploration of the common bile duct. In: Davenport M, Heaton ND, Superina R (eds). *Surgery of the Liver, Bile Ducts and Pancreas in Children*, 3rd ed. London: CRC Press; 2017: pp. 119–30.

Esposito C, Corcione F, Settimi A, et al. Twenty-five year experience with laparoscopic cholecystectomy in the pediatric population – from 10 mm clips to indocyanine green fluorescence technology: Long-term results and technical considerations. *J Laparoendosc Adv Surg Tech A* 2019; 29(9): 1185–91.

Fernández-Bautista B, Mata DP, Parente A, et al. First experience with fluorescence in pediatric laparoscopy. *European J Pediatr Surg Rep* 2019; 7: e43–e46. doi: 10.1055/s-0039-1692191.)

Ostlie DJ, Adibe OO, Juang D, et al. Single incision versus standard 4-port laparoscopic cholecystectomy: A prospective randomized trial. *J Pediatr Surg* 2013; 48: 209–14.

Short SS, Frykman PK, Nguyen N, et al. Laparoscopic common bile duct exploration in children is associated with decreased cost and length of stay: Results of a two-center analysis. *J Pediatr Surg* 2013; 48: 215–20

Strasberg SM, Brunt LM. Rationale and use of the critical view of safety in laparoscopic cholecystectomy. *J Am Coll Surg* 2010; 211: 132–8.

55

Surgery for biliary atresia: Open and laparoscopic

MARK DAVENPORT AND ATSUYUKI YAMATAKA

HISTORY

John Thomson from Edinburgh described an infant dying from cirrhosis secondary to "congenital biliary obstruction" in 1891. Surgical exploration was increasingly advocated from the 1930s and the terms "correctable" and "non-correctable" became prevalent to describe what could be done with a conventional surgical operation (e.g. hepaticojejunostomy). However, as most cases of biliary atresia (BA) were anatomically "non-correctable" at that time, their outlook was poor and true survivors were exceptional.

During the 1950s and 1960s Morio Kasai, a Japanese surgeon from Sendai, developed a more radical approach to the dissection, exposing residual microscopic bile ductules within the apparently solid parts of the porta hepatis. This operation (portoenterostomy) resulted in a much larger proportion of children who lost their jaundice following restoration of bile flow. Nevertheless, most still developed chronic liver disease. Liver transplantation became a practical option during the 1980s in the UK and North America and in the 1990s in Japan. Initially, this was for older children with life-threatening complications of liver disease (e.g. portal hypertension) but then it became available for infants of whatever age who simply had had no response to a Kasai operation or were deemed too far gone with established cirrhosis to benefit from one.

EPIDEMIOLOGY

Biliary atresia remains a rare disease with a frequency of between 1 in 10 000 and 1 in 20 000 live births. It is more common in Japan and China than in Europe or North America, although the reasons are not apparent. There is female preponderance for those with developmental BA (i.e. syndromic and cystic BA).

BA is a cholangiopathy of the extra- and intrahepatic parts of the biliary tree which, if untreated, leads to hepatic fibrosis and ultimately cirrhosis. The histological appearance of the liver is characterized by portal tract edema, bile duct plugging and proliferation, a small cell infiltrate, and variable amount of giant cell formation. Bridging fibrosis is a late feature in some cases, even at diagnosis.

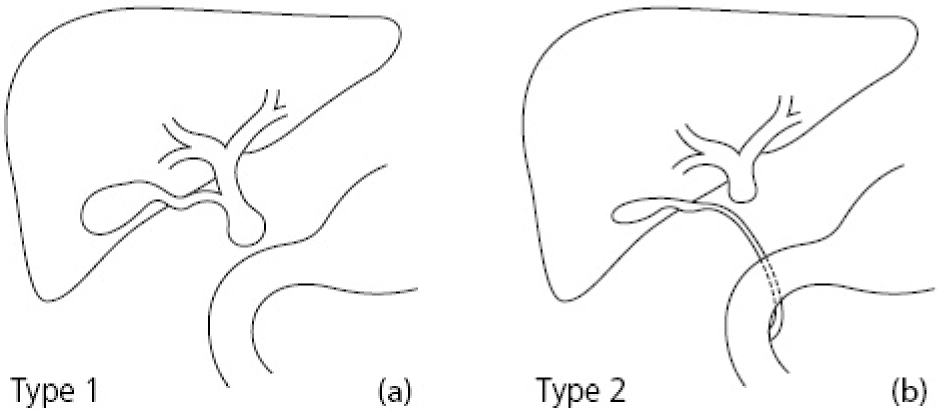
The lumen of the extrahepatic duct is obliterated at a variable level and this forms the basis for the commonest classification in use. Thus, there are three main types of BA:

- *Type 1 (~5%):* The level of obstruction is within the common bile duct (the gallbladder therefore contains bile).
- *Type 2 (~3%):* The level is within the common hepatic duct.
- *Type 3:* This is the commonest type (>90%), where there is no visible bile-containing proximal lumen and the obstruction is within the porta hepatis.

Cystic change can be seen in all types; some contain mucus, while others contain bile and this invariably causes diagnostic confusion with a choledochal cyst. In such cases, the wall is thickened and communicates poorly with abnormal non-dilated intrahepatic ducts, demonstrable on cholangiography.

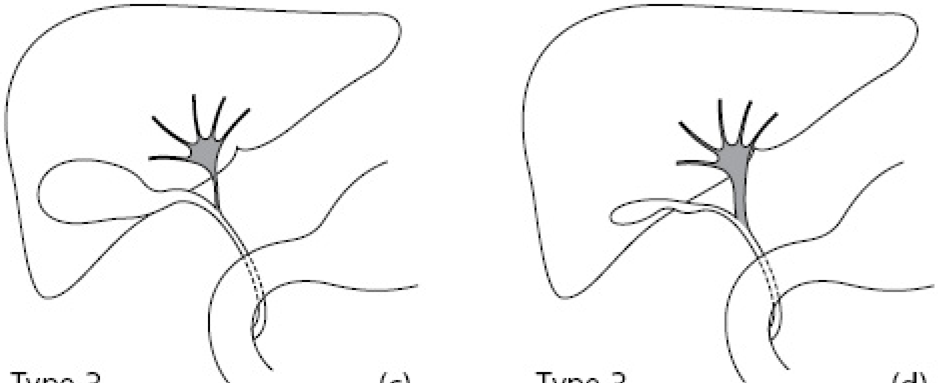
ETIOLOGICAL VARIANTS

There appear to be a number of variants of BA with differing etiology but the same phenotypic end point (**Figure 55.2**). For instance, there is a consistent association (~10% in European and North American series) with other non-biliary anomalies such as polysplenia, asplenia, cardiac malformation, situs inversus, absence of the inferior vena cava (IVC), preduodenal portal vein, and malrotation, for which the term “biliary atresia splenic malformation” has been used (**Figure 55.3a–c**). Such cases may result from some “insult” within the first trimester of pregnancy causing abnormal development of susceptible organ systems. Although such “insults” are hard to define, maternal diabetes, maternal thyrotoxicosis, and drug abuse have been reported. In about 5% of cases there is obvious cyst development within an otherwise obliterated biliary tree (cystic biliary atresia (CBA)) (**Figure 55.4a and b**); these are also highly likely to be developmental in origin as up to half may be detected on an antenatal ultrasound (US) scan, as far back as the sixteenth week of gestation. In most cases, however, only the biliary tree is abnormal and no obvious prenatal cause is identified (isolated biliary atresia). This has led to speculation that a perinatal hepatotropic virus infection (e.g. rotavirus, REOvirus, cytomegalovirus (CMV)) has been able to damage an otherwise normally developed biliary tree. Certainly, the group of infants who demonstrate IgM antibodies to cytomegalovirus can be distinguished clinically by having a more “hepatitic” histological appearance and more advanced degree of fibrosis in the liver with a reduced expectation of bile flow postoperatively (CMV IgM+ve BA). The recent discovery of a toxic substance (bilatresone) within some plant species may also contribute to an environmental cause in some cases though its relevance to humans is not known.



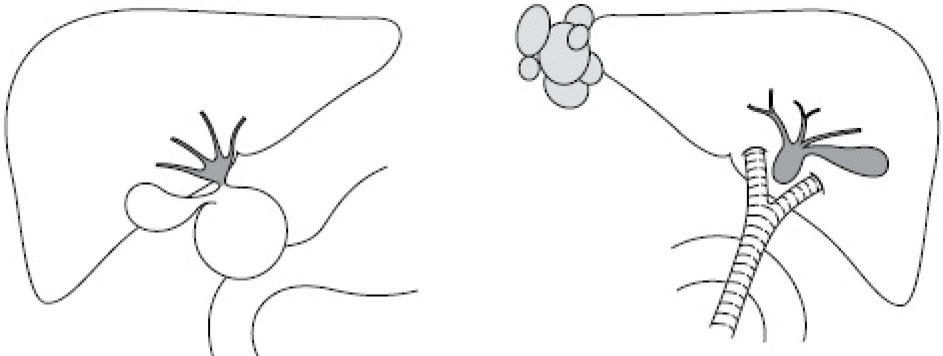
Type 1 (a)

Type 2 (b)



Type 3 (c)
(with mucocele of gallbladder)

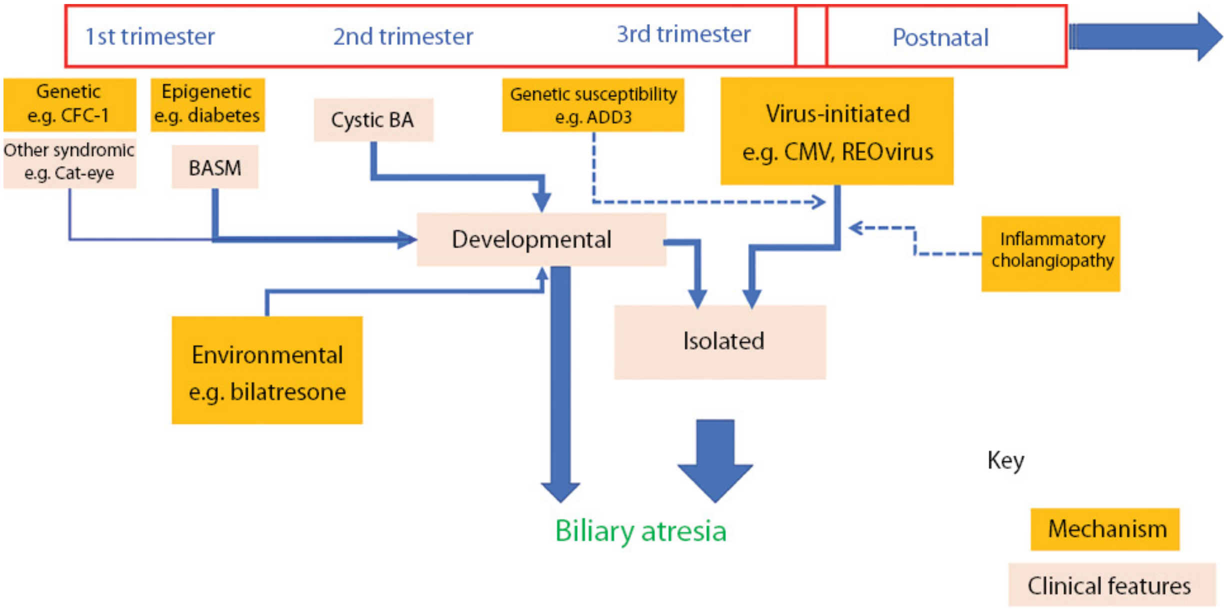
Type 3 (d)
(with atrophic gallbladder)



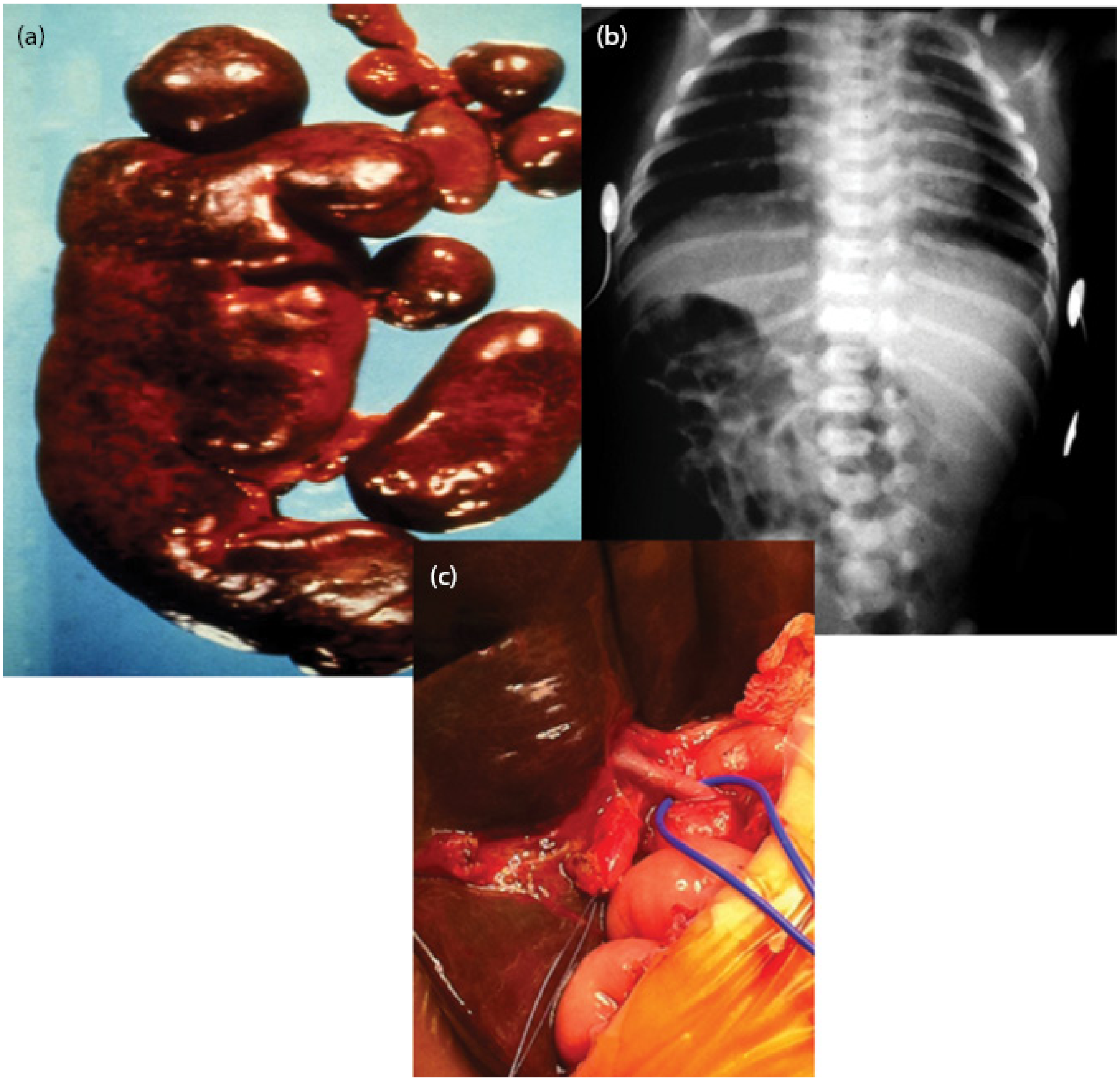
(e)
Cystic biliary atresia
(Type 3)

(f)
Biliary atresia splenic
malformation (with
preduodenal portal vein)

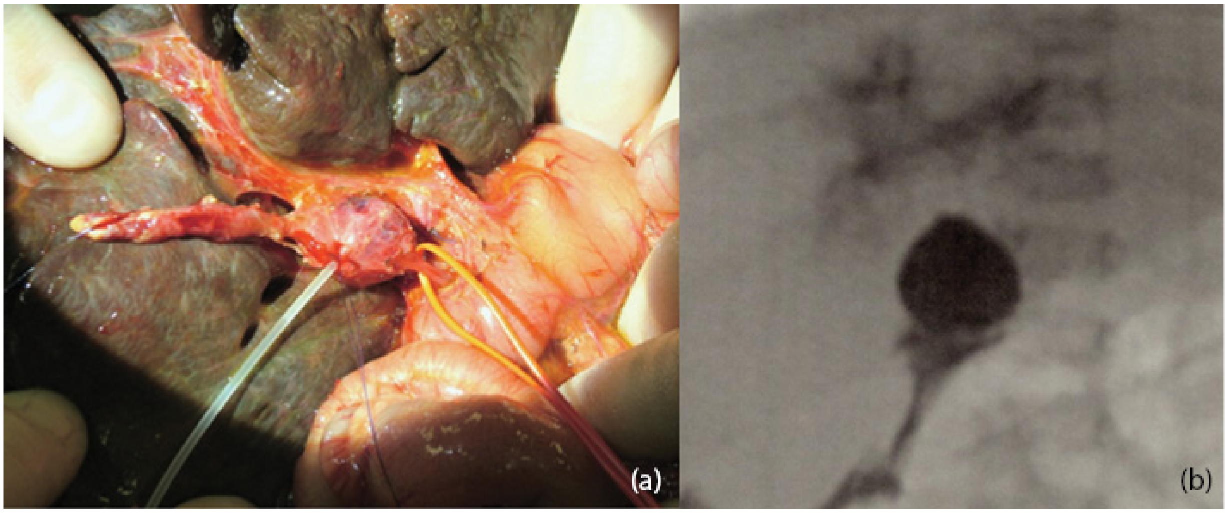
55.1a–f Classification of biliary atresia.


1st trimester
2nd trimester
3rd trimester
Postnatal
Genetic e.g. CFC-1
Epigenetic e.g. diabetes
Cystic BA
Genetic susceptibility e.g. ADD3
Virus-initiated e.g. CMV, REOvirus
Other syndromic e.g. Cat-eye
BASM
Developmental
Inflammatory cholangiopathy
Isolated
Environmental e.g. bilatresone
Key
Biliary atresia
Mechanism
Clinical features

55.2 Schematic representation of etiological heterogeneity.



55.3a–c Biliary atresia splenic malformation syndrome: Typically infants will have polysplenia (a), situs inversus (b), and a preduodenal portal vein (c). (Reproduced from Davenport et al., *Surgery of the Liver, Bile Ducts and Pancreas in Children*, 3rd ed., London: CRC Press; 2017, with permission.)



55.4a,b Cystic biliary atresia: (a) Operative image with cannulated cyst. Note the solid, atrophic gallbladder and string-like distal common bile duct. (b) Cholangiogram showing cyst and "cloud-like" appearance of the primitive intrahepatic ducts. (Reproduced from Davenport et al., *Surgery of the Liver, Bile Ducts and Pancreas in Children*, 3rd ed., London: CRC Press; 2017, with permission.)

PREOPERATIVE ASSESSMENT AND PREPARATION

All will be clinically jaundiced, and have pale stools and dark urine when looked for. This is due to failure of biliary excretion of conjugated (i.e. water-soluble) bilirubin into the gastrointestinal tract. Rising serum levels spill over into the urine causing its colour to darken. Such alternative pathways of bilirubin excretion are more developed (or perhaps better retained) in the neonate and very high levels of bilirubin (>300 µmol/L ≈ 17 mg/dL) is not a feature (as might be seen in adults with complete biliary obstruction).

Some infants with CBA will have had an abnormal antenatal maternal US scan and show a "subhepatic cyst."

Table 55.1 illustrates the differential diagnosis of conjugated jaundice in infants. It can be divided into principally medical and surgical causes, with the former predominating.

Table 55.1 Causes of conjugated jaundice in infancy

	Comments	Key diagnostic tests
Surgical		
Biliary atresia		US, liver biopsy, radio-isotope test, laparoscopy
Choledochal cyst	+ve antenatal scan	US, MRCP
Spontaneous perforation of bile duct	Usually unremarkable term infants	US, MRCP, radioisotope scan
Inspissated bile syndrome	Usually preterm and complicated neonatal history	US, MRCP, percutaneous transhepatic cholangiogram
Medical		
Alfa-1-antitrypsin deficiency		Serology, liver biopsy
Alagille's syndrome	Characteristic "elfin" facies, cardiac anomalies	Liver biopsy, echocardiogram, family history
Parenteral nutrition associated cholestasis	Preterm, prolonged gastrointestinal rest (e.g. NEC).	History, US, liver biopsy
TORCH infections	e.g. toxoplasmosis, CMV hepatitis	Serology, liver biopsy
Progressive familial intrahepatic cholestasis	Usually ↓ γGT, family history	Genetic screen, liver biopsy
"Neonatal hepatitis"	After exclusion of the above	Biochemistry, liver biopsy

The diagnostic workup should always include US scan; biochemical exclusion of α1-anti-trypsin deficiency and cystic fibrosis; and (in our institution) a percutaneous liver biopsy. Using this, about 80% of patients will have a positive diagnosis prior to laparotomy. Other techniques which have been described include endoscopic retrograde cholangiopancreatography (ERCP), percutaneous cholangiography, duodenal intubation, and measurement of bile. These may be useful (or, more likely, more practical) in selected cases and other centers. Currently, magnetic resonance cholangiopancreatography (MRCP) is not sufficiently detailed to confidently diagnosis BA, although it has shown promise for the more obvious structural problems such as a choledochal cyst. Radio-isotope hepatobiliary imaging (e.g. using iminodiacetic acid (IDA) derivatives) was formerly a popular investigation in distinguishing BA from neonatal hepatitis but can be difficult to interpret.

Most infants presenting within 70–80 days will not show clinical features of cirrhosis or irretrievable liver damage (e.g. gross ascites, nodularity on ultrasonography, or histological cirrhosis on liver biopsy) and should undergo laparotomy with the intention of performing a Kasai portoenterostomy (KPE). Others, who do show these features, may be considered for liver transplantation as a primary procedure. Nevertheless, it should be realized that prognosis is difficult to predict with any certainty and even a poorly functioning KPE may delay the need for a donor organ.

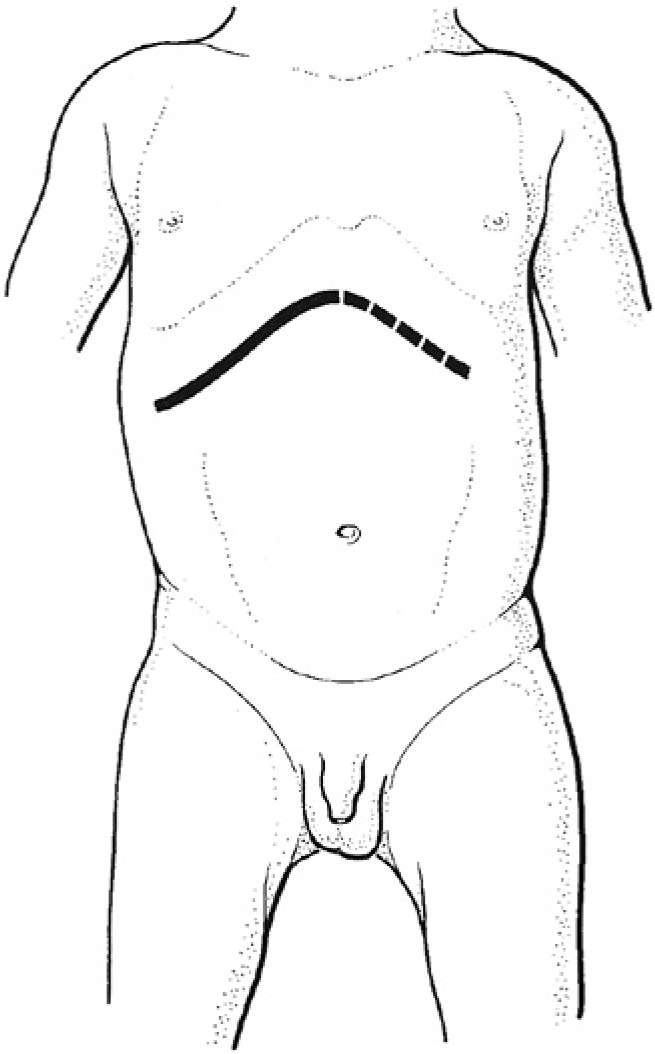
Blood investigations should include coagulation tests, such as international normalized ratio (INR), to exclude a vitamin K-dependent coagulopathy and all infants coming to surgery should have parenteral vitamin K (phytomenadione, 1 mg/day). The choice of parenteral antibiotics depends on local policies but should be broad-spectrum with reasonable bile penetration (e.g. second- and third-generation cephalosporins). In our institution these are given intravenously for 5 days and then orally for a further 25 days. Others continue these for up to 1 year.

OPEN KASAI PORTOENTEROSTOMY

The aim of open surgery is to excise all extrahepatic biliary remnants to allow a wide portoenterostomy reconstruction onto a portal plate, denuded of all tissue (**Video 55.1**). This should be the object not only in Type 3 BA but also in those who do have a visible bile-containing proximal communication (i.e. Types 1 and 2). It should be obvious therefore that frozen section, formerly used to confirm patent ductules, is not necessary because it should not be possible to resect any further biliary tissue. Attempts to excise parts of the liver, to "core out," or to "chase" the ductules have no value and are obsolete.

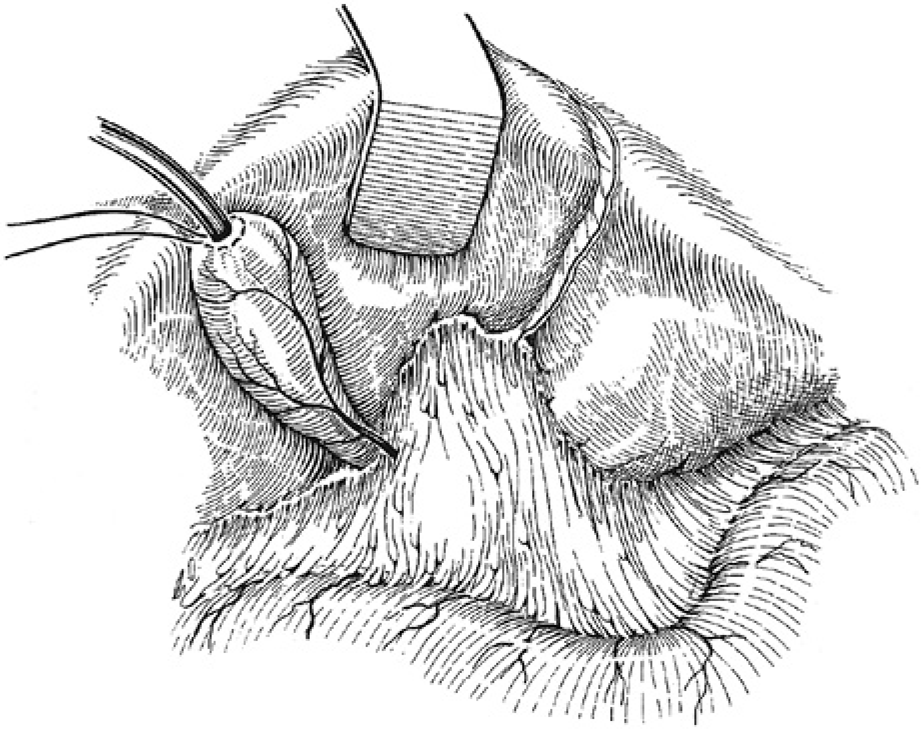
Incision

A short right (or left if situs inversus) upper quadrant muscle-cutting incision should be performed initially to confirm the suspected diagnosis or, if not immediately obvious, to perform an on-table cholangiogram (**Figure 55.5**). Laparoscopy may also achieve the same object at this stage. If there is no bile, or only clear mucus, in a collapsed, atrophic gallbladder, it is invariably BA. In those circumstances where there is obvious bile in the gallbladder, an on-table cholangiogram is clearly indicated—though in that situation BA is already excluded (apart from the rare cystic Type 1 BA, which should be obvious).



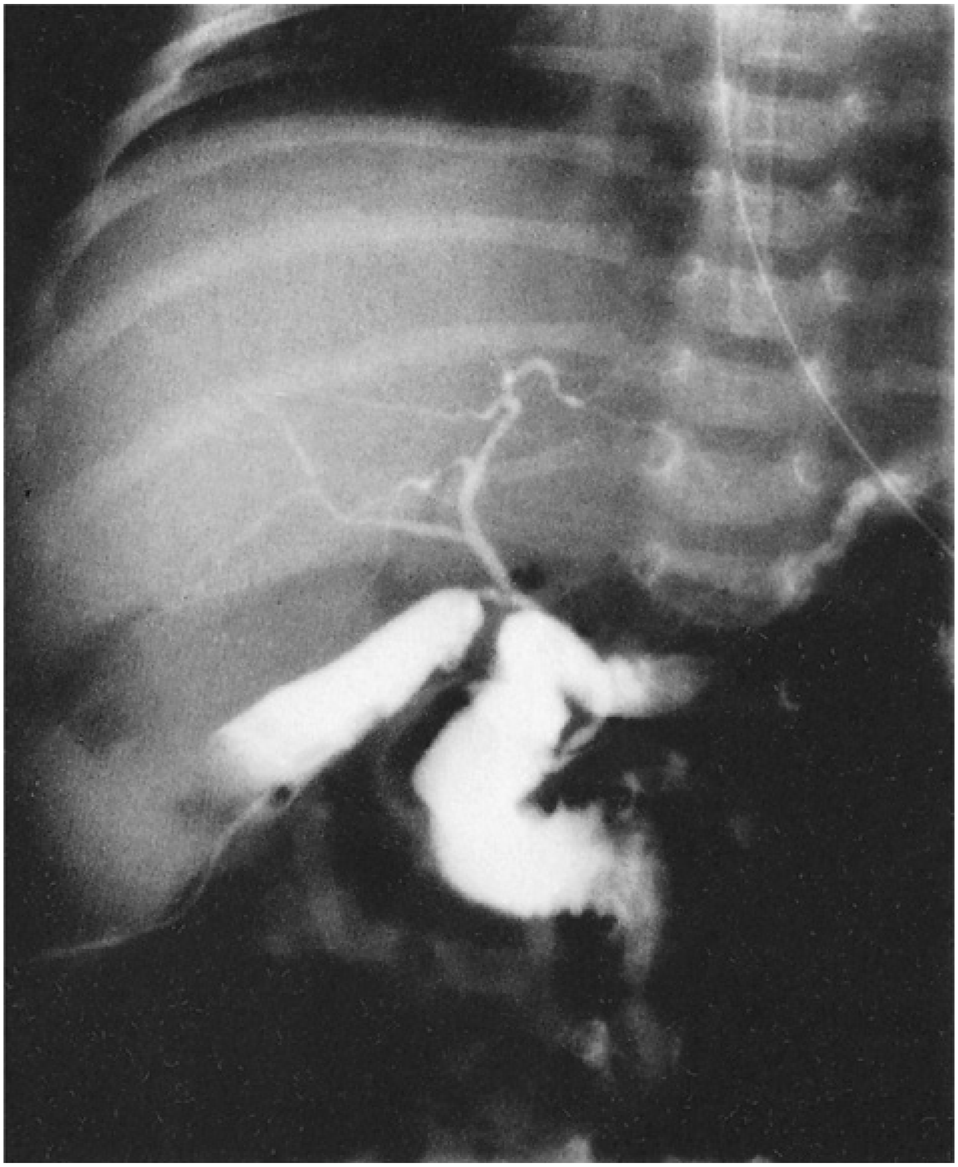
55.5

This is done by inserting a small feeding tube (4 Fr) into the gallbladder secured by a purse-string suture (**Figure 55.6**).



55.6

The demonstration of a patent common bile duct and intrahepatic ducts excludes a diagnosis of BA (**Figure 55.7**).



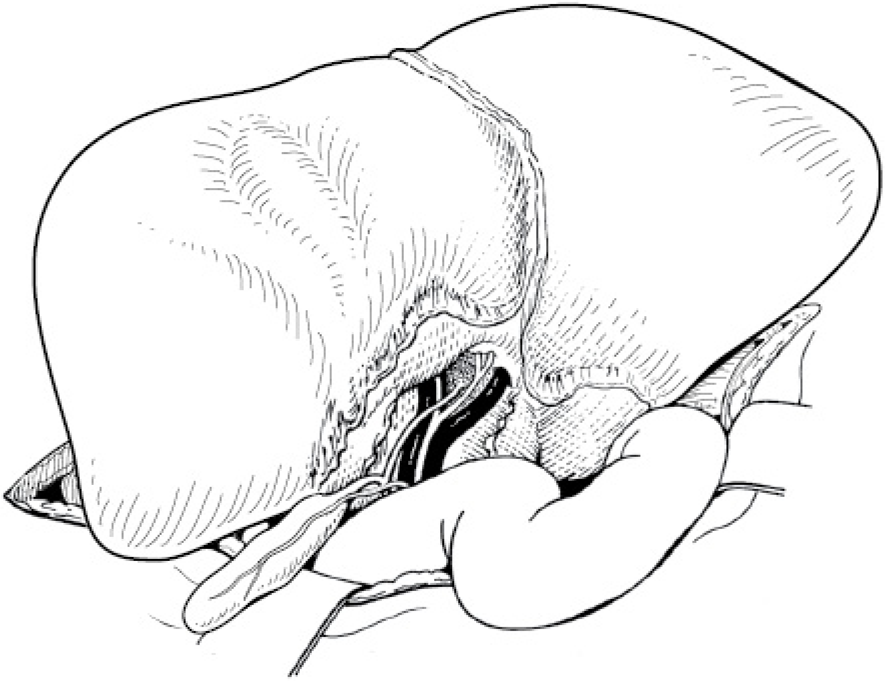
55.7

Sometimes, proximal passage of contrast into intrahepatic can be difficult to show as typically it preferentially fills only the distal duct and duodenum. This can be prevented by a small vascular or “bulldog” clamp on the common bile duct.

The incision can then be lengthened after confirmation of the diagnosis to cross the midline and ligate and divide the falciform ligament. The laparotomy should look carefully for other anomalies (e.g. polysplenia, preduodenal portal vein, absence of the IVC, malrotation) which may alter the subsequent technique.

Mobilization of the liver

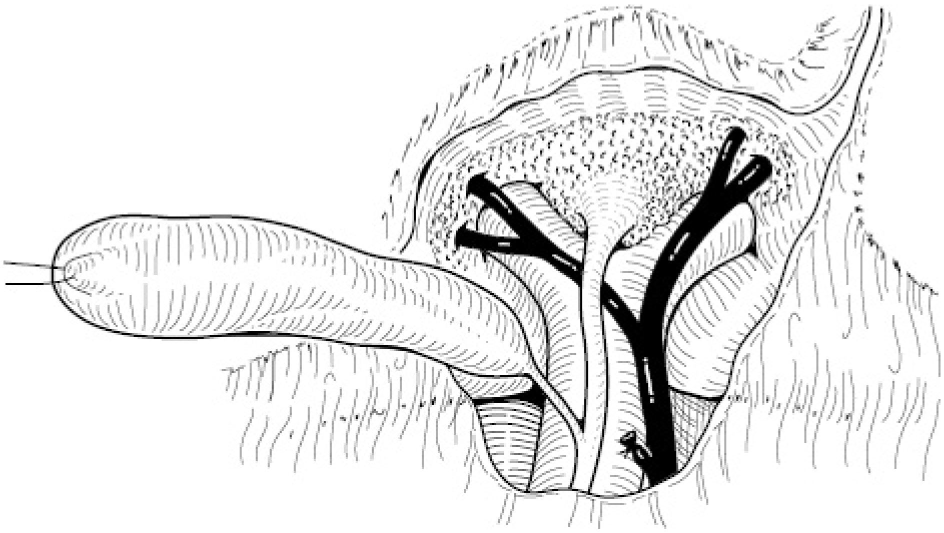
The liver should be mobilized and everted onto the abdominal wall (**Figure 55.8**). This can be achieved by dividing the falciform ligament and left coronary ligaments. The right-sided attachments around the "bare" area of the liver can be left intact and the liver still everted. The authors believe this to be a crucial step, which allows full exposure of the portal hepatis and facilitates the subsequent detailed dissection. Nonetheless, it is possible to dissect the porta with the liver *in situ* although most proponents will sling the portal vessels to achieve the visualization of the portal plate. It is imperative to warn the anesthetist at this stage as the maneuver impairs venous return to the heart by kinking the cava and will need an increase in IV volume support.



55.8

Mobilization of the gallbladder and bile ducts

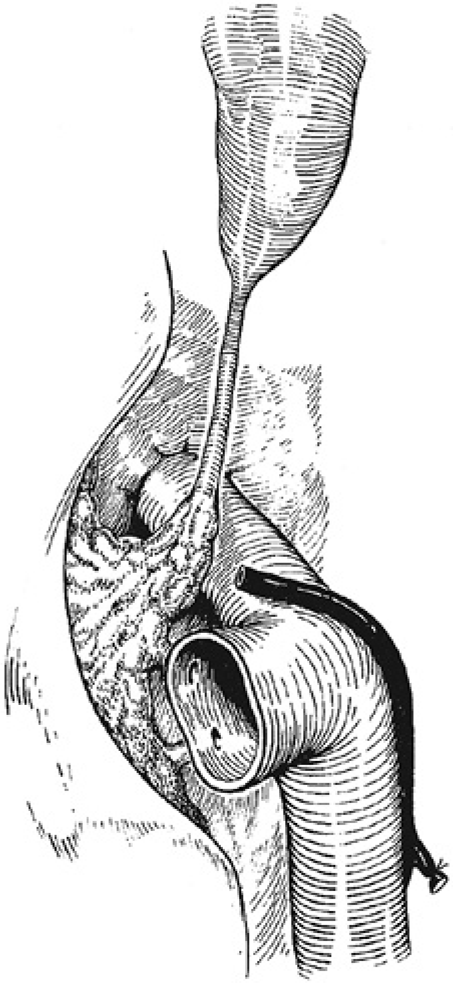
A stay suture on the gallbladder allows its elevation and separation from the bed with bipolar diathermy. The peritoneum overlying the portal triad (the hilar plate) is then divided and the various vascular (hepatic artery and portal vein) and biliary structures positively identified (**Figure 55.9**). Although parts of the biliary tract may be missing usually there is an intact biliary structure conforming to the usual arrangement of gallbladder, cystic duct, and common hepatic and common bile duct. The distal common bile duct is ligated and divided and the proximal part elevated from underlying connective and lymphatic tissue. Following the divided end of the cystic artery should lead to identification of the right hepatic artery, which may be superficial or deep to the common duct and may need slinging.



55.9

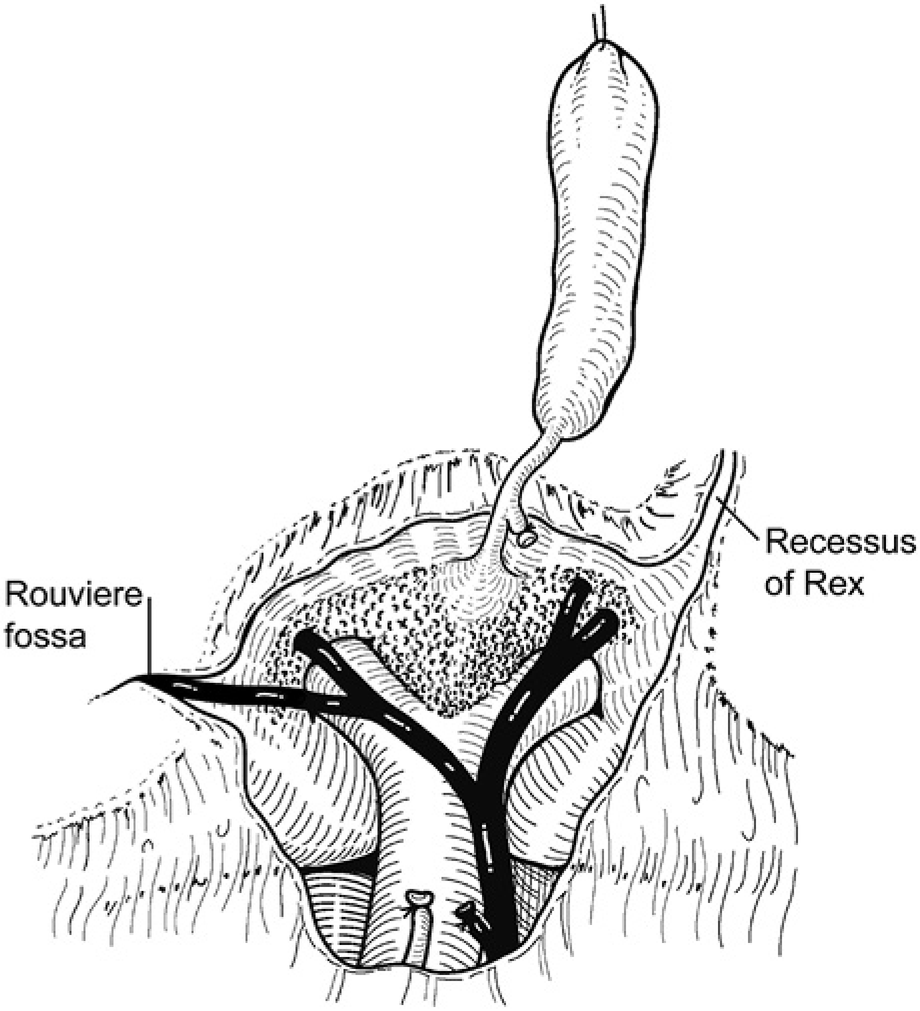
Dissection and exposure of the porta hepatis: Radical resection

Gradual separation of the biliary remnant will lead proximally to the porta and its relationship to the bifurcation of the portal vein will become clearer (**Figure 55.10**). The bifurcation is gradually freed from the porta, typically by division of small veins crossing into the central portion. In the posterior part of this, the caudate lobe should become visible.



55.10

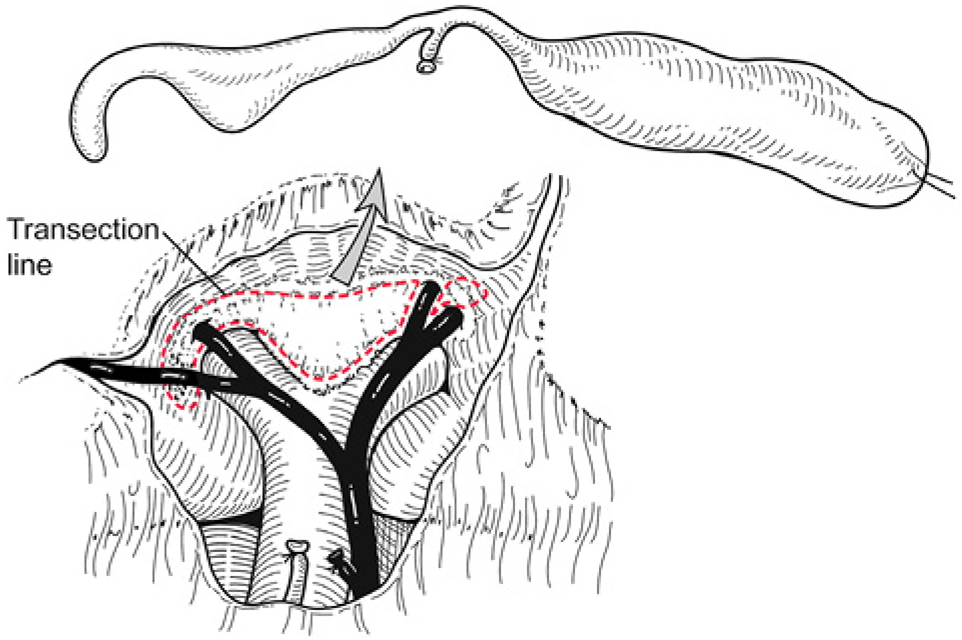
The dissection is then extended to both right and left aspects. On the left, consideration should be given to fully exposing the so-called "recessus of Rex" by dissecting the umbilical vein down to its origin from the left portal vein (**Figure 55.11**). An isthmus of liver parenchyma (from segment 3 to 4) may need division by coagulation diathermy to achieve this. This then becomes the proximal extent of the left-sided dissection. On the right side, remnant biliary tissue can be identified around the right vascular pedicle encompassing not only the base of the gallbladder fossa but also at the extreme a small fossa (the Rouviere fossa) containing posterior branches of right hepatic vessels. Although some authors advocate slinging the right and left portal veins, this may compromise subsequent liver blood flow and simple judicious retraction should give more than adequate exposure.



55.11

Excision of biliary remnants

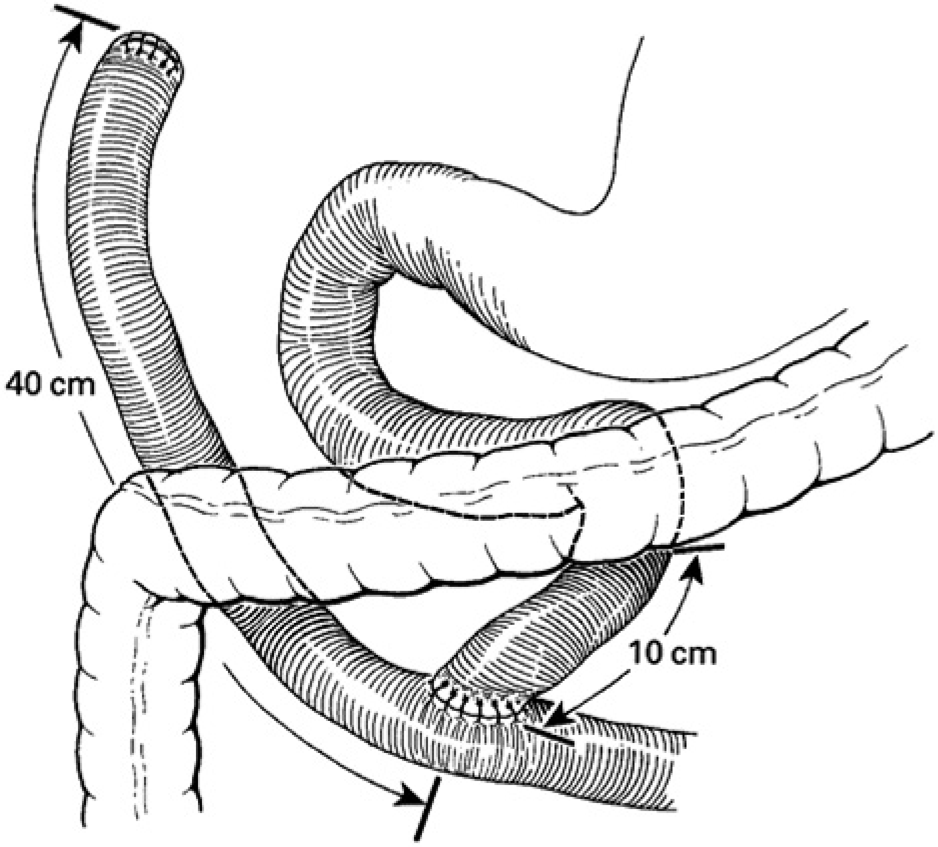
The remnant can now be removed (**Figure 55.12**). Beginning at the right side and using a combination of small curved sharp tenotomy scissors and/or scalpel, a plane is developed between solid white biliary remnant and the underlying liver, absolutely flush with the capsule. The appearance at the transected portal plate should be almost translucent, denuded of tissue, but with liver tissue not actually transgressed. Excising liver parenchyma does not improve bile drainage, presumably because any divided ductules simply become obliterated by subsequent scar tissue. Untoward attempts at hemostasis of the porta by diathermy at this stage should be avoided. The area should be packed and the liver returned to the abdominal cavity, which will relieve venous congestion.



55.12

Formation of Roux loop

The duodenojejunal junction should be identified and a point about 10 cm from this should be mobilized to be the site of the jejunal anastomosis. Bowel is divided using a linear stapler, and a Roux loop, measured along the antimesenteric border at approximately 40 cm, constructed (**Figure 55.13**). This loop can then be brought through a right-sided window in the transverse mesocolon. An end-to-side jejunal anastomosis is constructed using whatever technique the operator is comfortable with (e.g. side-side stapling using Endo GIA□ or handsewn end-to-side using 5/0 PDS□ sutures).

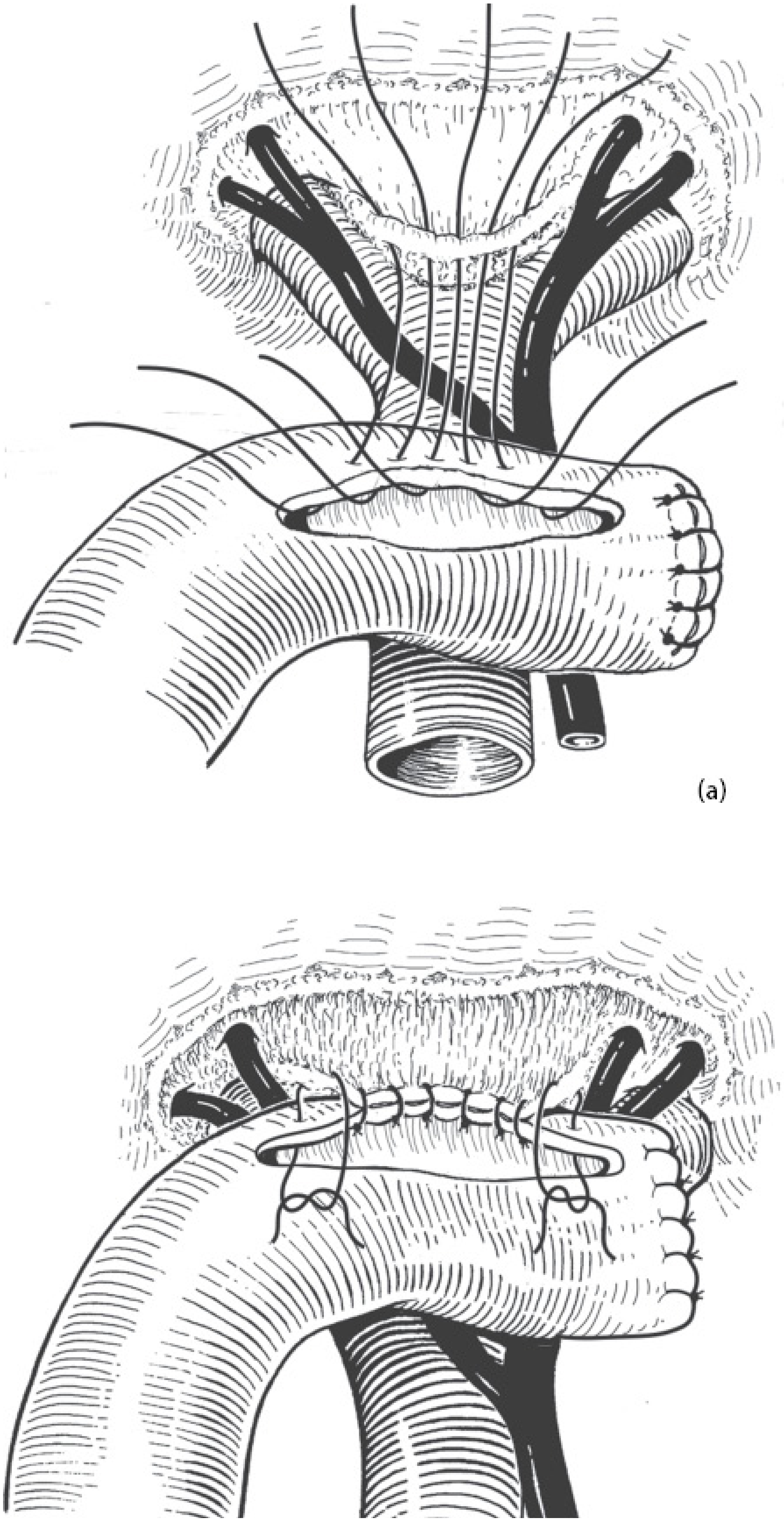


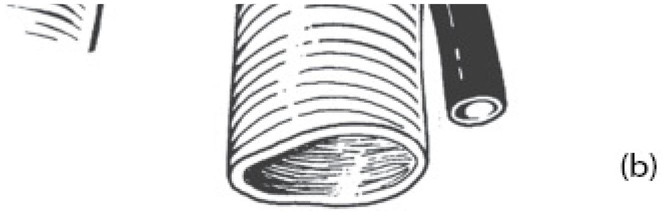
55.13

The end of the Roux loop should have enough length to reach the porta without undue tension.

Portoenterostomy

The liver is again everted to outside the abdominal cavity (**Figure 55.14a**). A non-crushing bowel clamp placed along the jejunal loop keeps this part steady and an opening is made in the antimesenteric aspect about 0.5 cm from the stapled end.


(a)



55.14a,b

It is easier if the posterior row of sutures (e.g. 6/0 PDS®) is placed sequentially and held with mosquito clips until the row is complete. The jejunal loop can then be “parachuted” down to the porta and the sutures tied (**Figure 55.14b**). The anterior row is then completed. The anastomosis should be wide, measuring about 2 cm, if there has been an adequate dissection of the right and left sides.

The mesocolic window around the Roux loop is closed, avoiding any kinking. A drain can be left, particularly if there has been more than a small quantity of ascites, but this is not essential. The wound closure, though, needs to be secure and watertight.

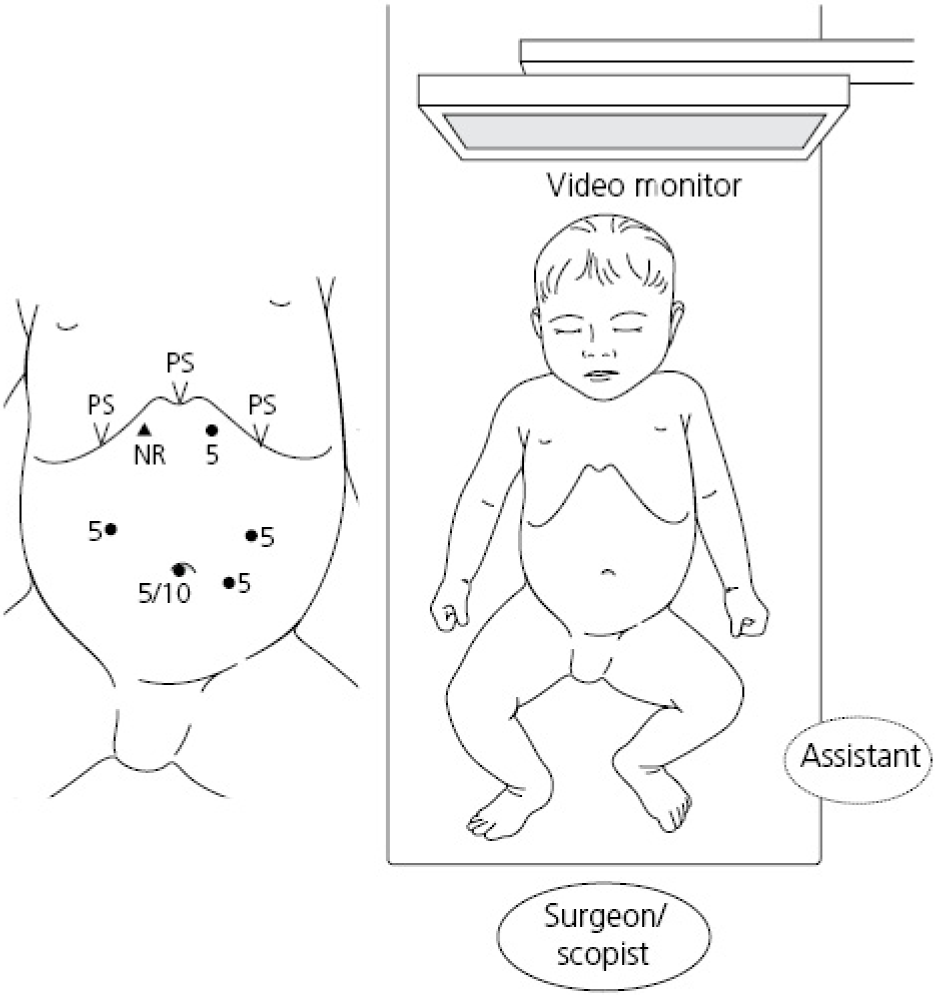
Surgical alternatives

There have been many alternatives to the original description of the KPE. The crucial part (viz. the portal dissection) has certainly become more extensive and widened on both right and left sides, as described here in the open operation, although this is retained for the laparoscopic operation. Most alternatives have included variations of the Roux loop, involving the creation of either stomas or interposed “antireflux” valves. The aim of these was to reduce postoperative cholangitis. None has stood the test of time, however, and a straightforward Roux loop reconstruction is all that is required.

LAPAROSCOPIC KASAI PORTOENTEROSTOMY

Positioning

The infant is positioned at the foot of the table, with the surgeon at the patient's feet. A 300 5 mm or 10 mm laparoscope is used, inserted through a 10 mm trocar placed supraumbilically using an open Hasson technique (**Figure 55.15**). Three additional ports are used: Two 5 mm ports on either side of the midline for the operator's right and left hands, just lateral to the rectus. A final port is inserted below the umbilicus on the left. A Nathanson retractor (marked NR on **Figure 55.15**) (Teleflex Medical, High Wycombe, UK), placed through the epigastrium, is crucial to expose the porta for the anastomosis. Finally, percutaneous stay-sutures (PS) are also used to expose and elevate the liver margin by ensnaring the umbilical ligament, and right and left lobes.

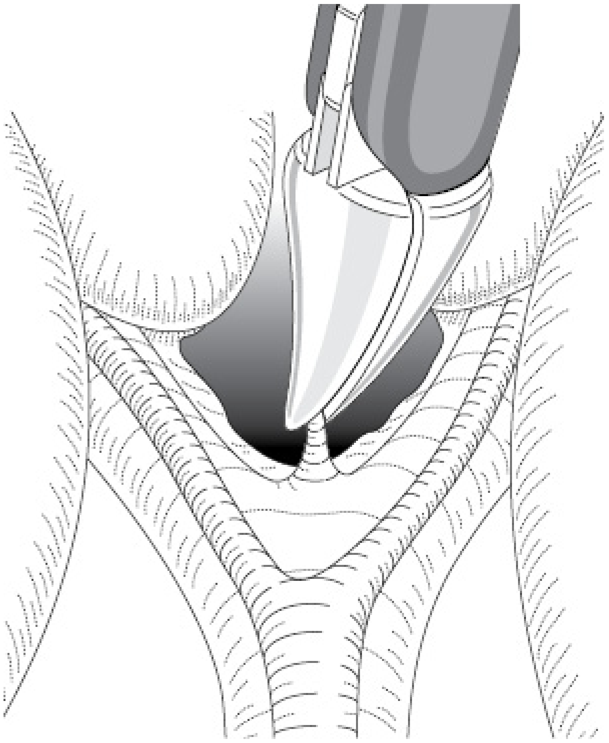


55.15

Gallbladder and porta hepatis dissection

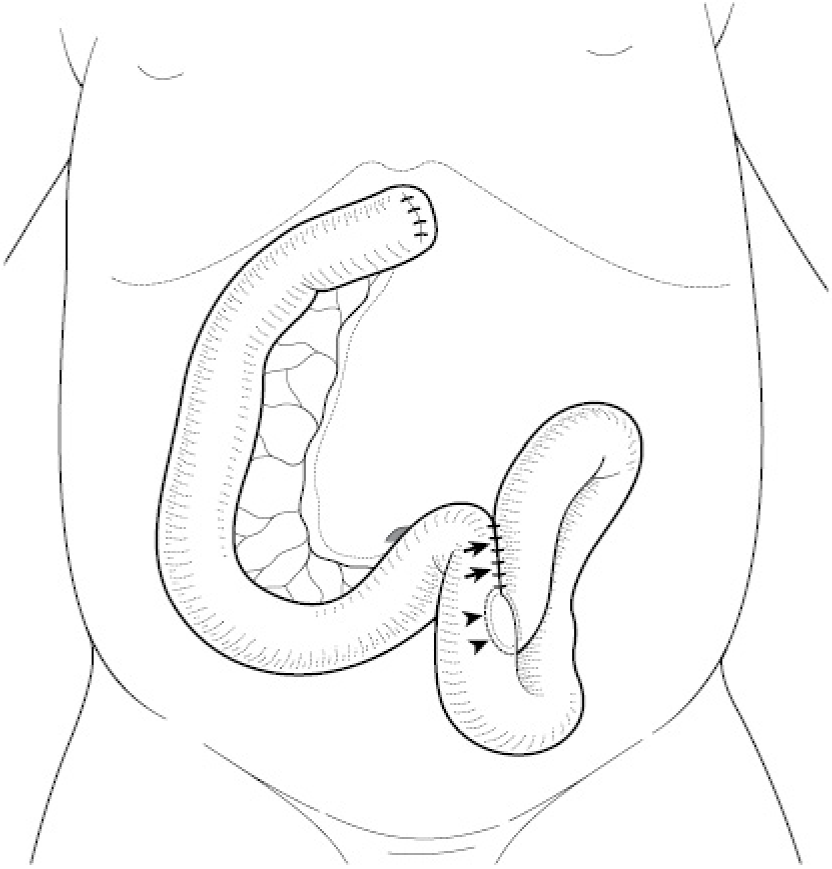
Hook diathermy and tissue forceps are used to dissect the cystic duct and mobilize the gallbladder and distal bile from the duodenum.

A further 5 mm trocar is placed in the epigastrium specifically for a LigaSure□ device (Valleylab, Boulder, CO, USA) (**Figure 55.16**). This is used to dissect the proximal biliary remnant, especially after dividing small venules from the portal vein into the remnant, and it minimizes thermal injury to bile ductules in the cone of biliary remnant tissue during dissection. The level of transection is more akin to Kasai's original description than the current extended variant described previously.



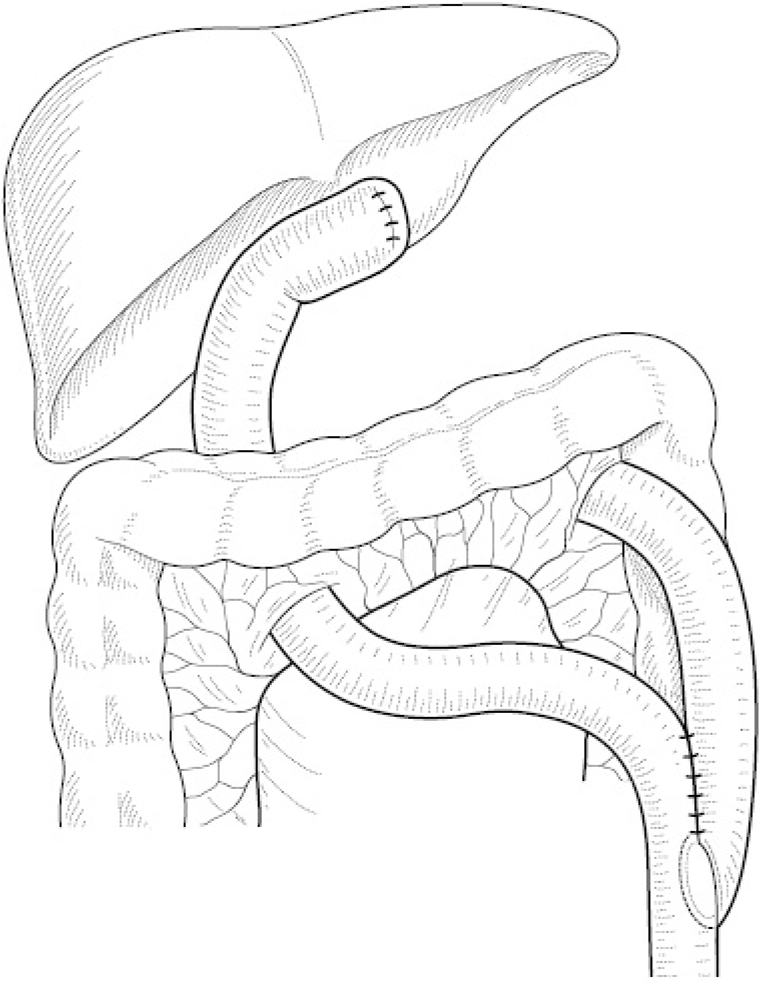
55.16

The ligament of Treitz is identified and the proximal jejunum about 15 cm from this identified and exteriorized through the umbilicus. This is then divided and the length of the Roux loop calculated by bringing it to the xiphoid process on the anterior abdominal wall (i.e. length not actually predetermined as 30 cm or 40 cm, etc.) (**Figure 55.17**).



55.17

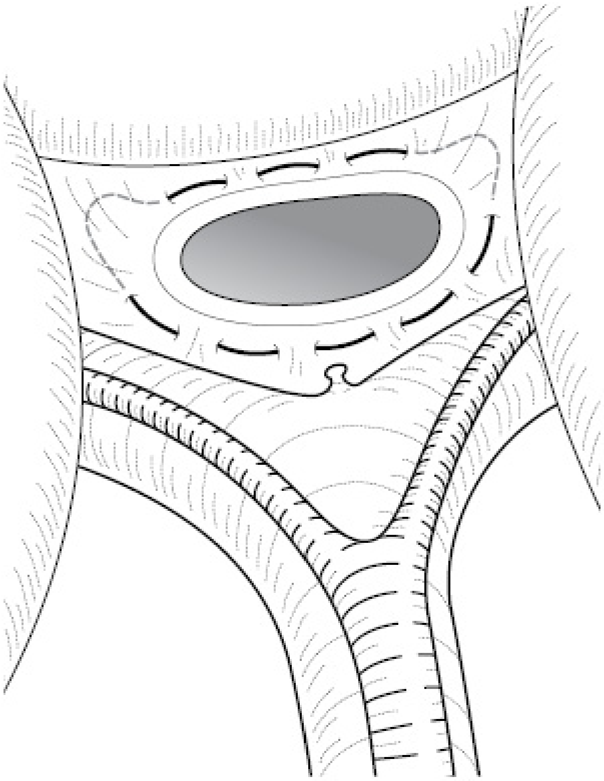
The “short” Roux limb is approximated to the native jejunum into the Roux loop such that the jejunojejunostomy will fit naturally into the splenic flexure (**Figures 55.18** and **55.19**). Finally, an antimesenteric enterotomy 10 mm in length is made near the close end and the whole returned to the abdominal cavity. This is passed through the mesocolon to lie without tension at the porta hepatis.



55.18

Portoenterostomy

Anastomotic sutures (5/0 or 6/0 PDS) are placed between the enterotomy and the liver parenchyma around the margins of the transected portal plate (**Figure 55.19**). These should be deep enough to prevent leakage but shallow enough to avoid residual ductules. At the 2 o'clock and 10 o'clock positions no sutures are placed to avoid damaging any recapitulated ducts in these positions. Sutures may be placed away from the plate into adjacent perivascular adventitial tissue.



55.19

Surgical philosophy in the porta hepatis

Many alternatives have been proposed since Kasai's original description, with the drift being to a more radical dissection aiming to excise all extrahepatic bile duct tissue within and between the vascular pedicles and incorporating this into a wide anastomosis. The laparoscopic dissection described here is deliberately abbreviated to transecting the cone leaving an ovoid remnant.

POSTOPERATIVE CARE

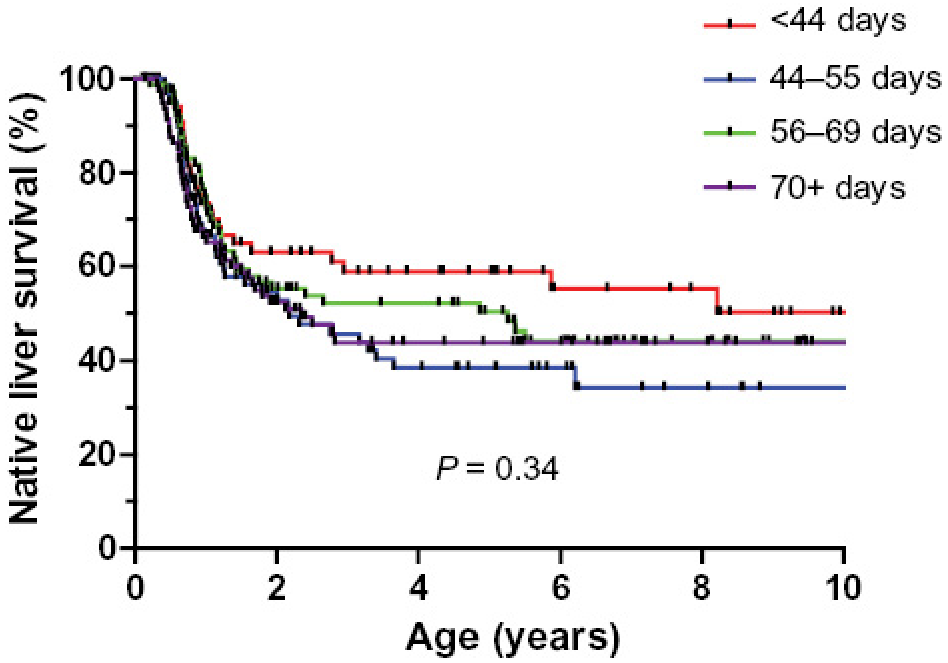
Intravenous fluids and nasogastric aspiration are continued until return of bowel function (after 3–4 days). Careful monitoring of blood glucose, electrolytes, and INR is important in the early phase. By about the fourth week there should be a definite fall in bilirubin and consistently pigmented stools in those who will do well.

High-dose corticosteroids (e.g. starting dose oral prednisolone 5 mg/kg/day; reducing in stages to 1 mg/kg/day over 4–5 weeks) and ursodeoxycholic acid (25 mg/kg/day) is a reasonable adjuvant regimen with a degree of evidential back-up and remains widely prescribed in Europe, Japan, and China. More controversial remains the treatment of those infants who are CMV IgM +ve. In our institution these infants now also undergo treatment with oral valganciclovir for a period, guided by the levels of CMV in the blood.

Strict attention to nutritional needs is important and all infants need regular vitamin supplementation. Medium-chain triglyceride (MCT) formula milk (e.g. Caprilon; SHS, Liverpool, UK) is advocated to maximize calorie input and facilitate lipid absorption.

OUTCOME

There are many factors which will influence surgical outcome in BA. Some are unalterable (e.g. degree of cirrhosis at presentation; absence, or paucity of, bile ductules at the level of section) but some may be subject to change (e.g. surgical experience, untreated cholangitis). The effect of age at surgery is complex and, while the principle that increasing age leads to worse outcome is unarguable, it can be difficult to discern and prove even in large series (**Figure 55.20**). Currently, in our centralized England and Wales experience involving three large centers with experienced surgeons, >65% of all infants undergoing KPE will clear their jaundice and achieve a normal (<20 μmol/L) bilirubin. These patients should do well and have a good quality of long-term survival with their native liver. In those with no effect from the KPE (usually apparent within 2–3 months), active consideration should be given to early liver transplantation.



55.20 Effect of age at Kasai portoenterostomy, England and Wales data. Infants with isolated biliary atresia ($n = 318$) were divided by age at surgery. There was no difference overall ($\chi^2 = 3.3$; $P = 0.34$) or for trend ($\chi^2 = 0.87$; $P = 0.35$). Specifically, there was no difference between the two outermost curves ($\chi^2 = 2.1$; $P = 0.15$). (Reproduced from Davenport et al., Biliary atresia in England and Wales: Results of centralization and new benchmark. *J Pediatr Surg* 2011; 46: 1689–94, with permission.)

The two principle complications which can be life-threatening are cholangitis and the development of esophageal varices due to portal hypertension.

- *Cholangitis:* This occurs most commonly in the year following primary surgery in about 40–50% of children. Paradoxically, it only occurs in children with some degree of bile flow, not in those with early failure. Clinically, it is characterized by worsening jaundice, fever, and acholic stools. The diagnosis may be confirmed by blood culture or by percutaneous liver biopsy but it is important to treat cases *on suspicion only* without waiting for confirmation. This should be with IV broad-spectrum antibiotics effective against Gram-negative organisms.
- *Portal hypertension:* Increased portal venous pressure has been shown in nearly all infants at the time of the KPE, although subsequent portal

hypertension depends on both the degree of established fibrosis and, most importantly, the response to surgery. There is a relationship with biochemical liver function and variceal development and, in those who fail and need early transplantation, about 30% will have had a significant variceal bleed. In those who respond well to initial KPE but who have established fibrosis, variceal development can be delayed with presentation with bleeding perhaps only occurring at 2–3 years. Current treatment of varices includes both injection sclerotherapy in the younger ones and variceal banding in those able to tolerate the endoscopic banding attachment.

VIDEO

OPS_8e_video_55.1 Kasai portoenterostomy

FURTHER READING

Davenport M. Biliary atresia: From Australia to the zebrafish. *J Pediatr Surg* 2015; 51: 200–5.

Davenport M, Heaton N, Superina R (eds). *Surgery of the Liver, Bile Ducts and Pancreas in Children*, 3rd ed. London: CRC Press; 2017.

Davenport M, Ong E, Sharif K, et al. Biliary atresia in England and Wales: Results of centralization and new benchmark. *J Pediatr Surg* 2011; 46: 1689–94.

Hartley J, Davenport M, Kelly D. Biliary atresia – review. *Lancet* 2009; 374: 1704–13.

Koga H, Miyano G, Takahashi T, et al. Laparoscopic portoenterostomy for uncorrectable biliary atresia using Kasai's original technique. *J Laparoendosc Adv Surg Tech A* 2011; 21: 291–4.

Wada M, Nakamura H, Koga H, et al. Experience of treating biliary atresia with three types of portoenterostomy at a single institution: Extended, modified Kasai, and laparoscopic modified Kasai. *Pediatr Surg Int* 2014; 30: 863–70.

56

Choledochal malformation

MARK DAVENPORT AND NGUYEN THANH LIEM

HISTORY

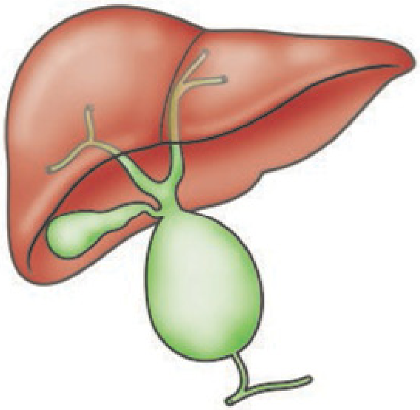
As well as describing the nature of the termination of bile and pancreatic ducts into the duodenum, the 18th-century anatomist Abraham Vater also described a choledochal cyst evident in one of his dissections. In 1852, Andrew Halliday Douglas, practising in Edinburgh, described a 17-year-old girl who had become deeply jaundiced with a palpable right upper quadrant mass. He attempted to treat this by trocar drainage with some success, though she ultimately died.

The first description of surgical excision of a cyst was in 1924 by the American surgeon, Golder McWhorter, who used a hepaticoduodenostomy as the mode of biliary reconstruction in a 49-year-old woman. The first successful laparoscopic excision and reconstruction was reported in a 6-year-old child in 1995 by an Italian team.

PRINCIPLES AND JUSTIFICATION

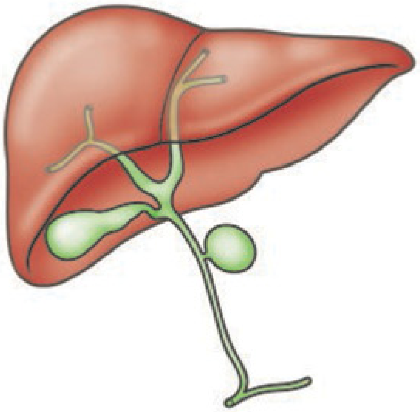
Although the traditional term “choledochal cyst” is still frequently overused, irrespective of the actual choledochal morphology and shape, this practice should be avoided. Congenital choledochal malformation (CCM) is a better term for the spectrum of biliary abnormalities characterized by dilatation. The incidence of CCM is not known though it is clearly much more common in East Asian countries and may even be increasing. All the common types have a female predominance.

The classification shown in **Figure 56.1** is the authors' modification of Todani's classification. Only Types 1 and 4 are commonly seen in children. Both types can be divided into cystic or fusiform depending on the character of the extrahepatic dilatation (hence Types 1c, 1f, 4c, and 4f). All four subtypes are usually associated with a varying degree of common pancreatobiliary channel, although the amount of pancreatic fluid reflux into the bile duct varies enormously.



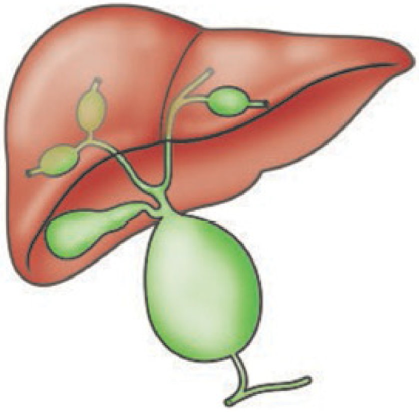
Type 1c

(a)



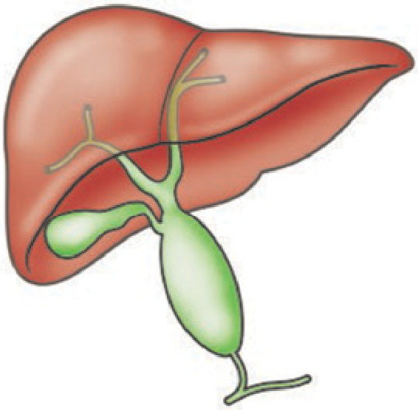
Type 2
(diverticulum)

(b)



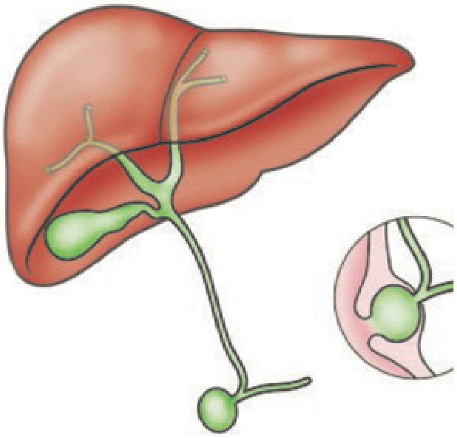
Type 4
(4c if cystic, 4f if fusiform)

(c)



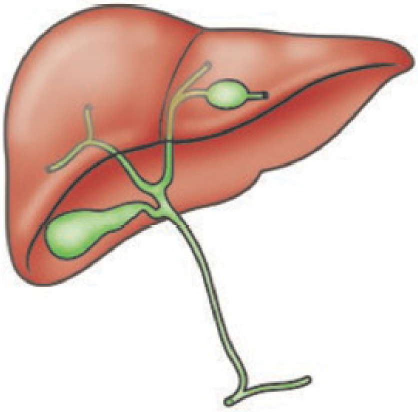
Type 1f

(d)



Type 3
(choledochocele)

(e)



Type 5
(isolated or with

(f)

Types 2 and 3 are rarely seen in children and may not even be congenital in origin. Type 5 malformations can be divided into those simply with isolated intrahepatic biliary dilatation and those with other features of Caroli's disease (or syndrome). This latter condition is invariably due to genetic mutation and would include bilateral renal cysts, failure or fibrosis, and early-onset liver fibrosis. Biliary dilatation in these children is characterized by multiple, discrete saccular lesions. They have a normal common bile duct (CBD) and no common channel.

It is possible to have a significant common channel with only minimal choledochal dilatation (formerly termed *forme fruste* choledochal cyst). The term "common channel syndrome" is probably better, and some of these children will present with recurrent pancreatitis, particularly if the common channel is dilated and filled with debris or stones.

CLINICAL FEATURES

Antenatal detection of Type 1c CCM is possible and needs to be urgently distinguished from cystic biliary atresia in the postnatal period. Other differential diagnoses might include parenchymal liver cysts and duodenal duplication.

The classical clinical trial has been jaundice and fever occurring in a child with a palpable right upper quadrant abdominal mass. This is now uncommon, perhaps occurring in only 10–15% of cases. Most cystic lesions present during early childhood with obstructive jaundice and acholic stools, depending on the degree of obstruction. Fusiform CCM, by contrast, tends to present later, often with recurrent abdominal pain, and sometimes with overt pancreatitis. If symptoms are disregarded, biliary cirrhosis may develop, though this is uncommon. Choledochal perforation may occur in a few, either directly into the peritoneal cavity where it can resemble acute-onset ascites or retroperitoneally tracking down the paracolic gutter and mimicking appendicitis.

An accurate diagnosis of CCM should be established preoperatively in virtually all cases. The current standard is ultrasound (US) followed by magnetic resonance cholangiopancreatography (MRCP) (ideally obtained using a 3.0 T magnetic resonance scan) and this should define most elements of key biliary and pancreatic anatomy. The degree of common duct and intrahepatic dilatation can be measured precisely, together with the presence or absence of luminal content. Defining the degree and dilatation of the common channel is more difficult but can be deferred to the operative cholangiogram.

Endoscopic retrograde cholangiopancreatography (ERCP) may still be needed for diagnosis in some children with suspected common channel syndrome presenting with recurrent pancreatitis and only borderline biliary dilatation. Occasionally, ERCP and preoperative stenting may need to be carried out for obstructed CM in older children.

All patients require biochemical liver function tests, amylase levels, and exclusion of coagulopathy by international normalized ratio (INR) testing.

SURGICAL MANAGEMENT

The principle of surgery is to excise the abnormally dilated part of the extrahepatic duct, typically to the level of the common hepatic duct (CHD) and down to the level of the junction with the common channel, within the head of pancreas with subsequent biliary reconstruction. This type of reconstruction has become controversial with the advent of a laparoscopic alternative. Though the standard remains that of hepaticojejunostomy en-Roux (named after Cesar Roux (1837–1934), a Swiss surgeon working mainly in Lausanne, who developed this for drainage of the obstructed stomach and not for the bile duct) for many, hepaticoduodenostomy is now acceptable.

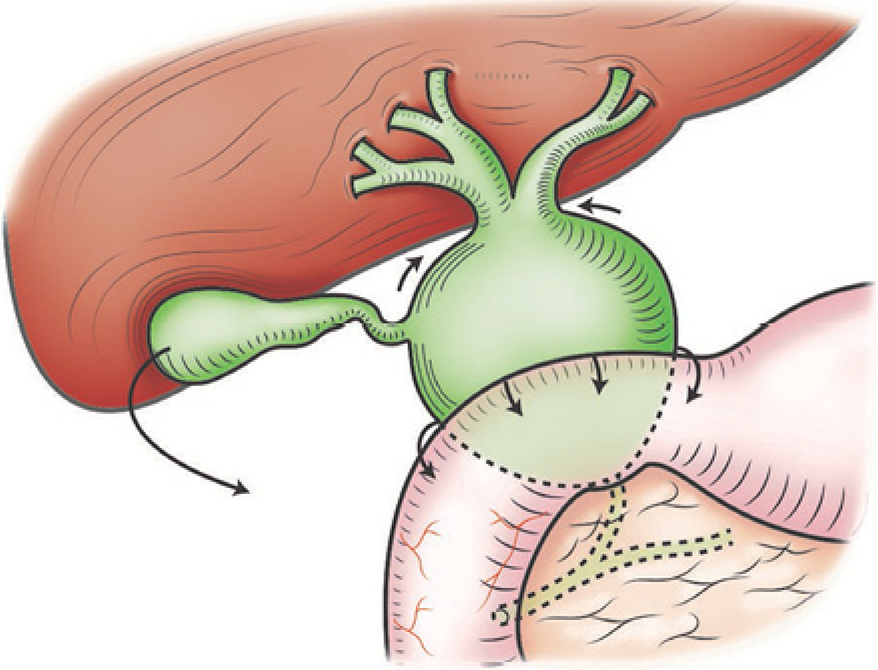
Early operations such as simple drainage into adjacent viscera (e.g. cystduodenostomy) are now obsolete as that procedure leaves the abnormal bile duct *in situ* to cause problems later on.

Open excision and biliary reconstruction

A right-sided, muscle-cutting subcostal incision gives appropriate access.

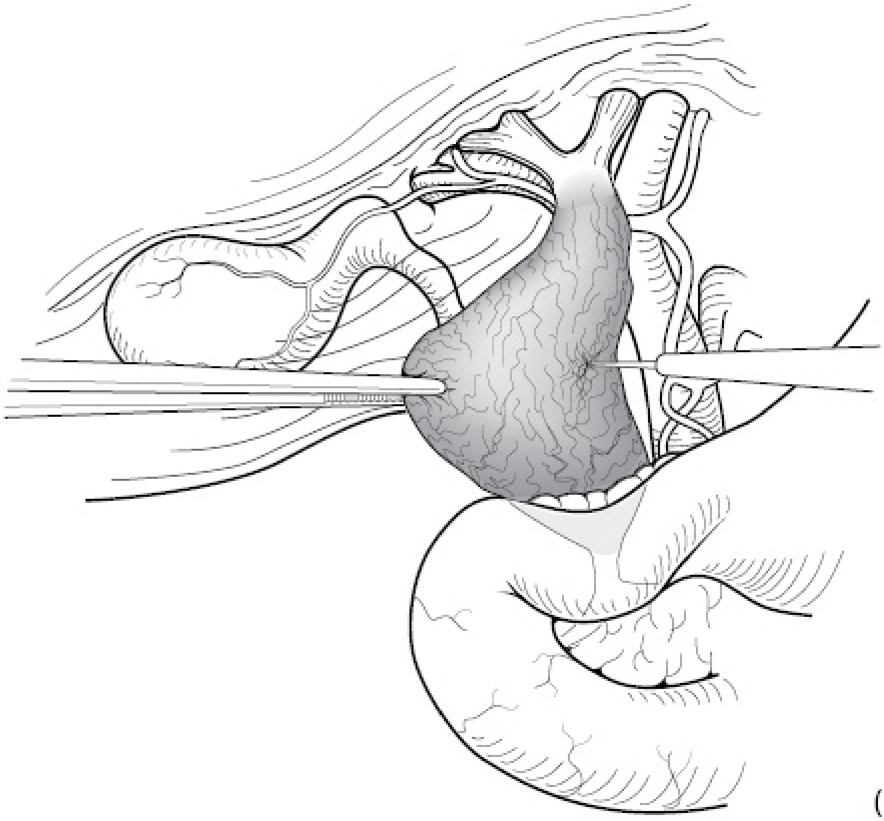
The liver should be assessed for any liver fibrosis or overt cirrhosis. Severe portal hypertension may make subsequent dissection difficult and dangerous and a period of external biliary drainage may be a better temporising solution.

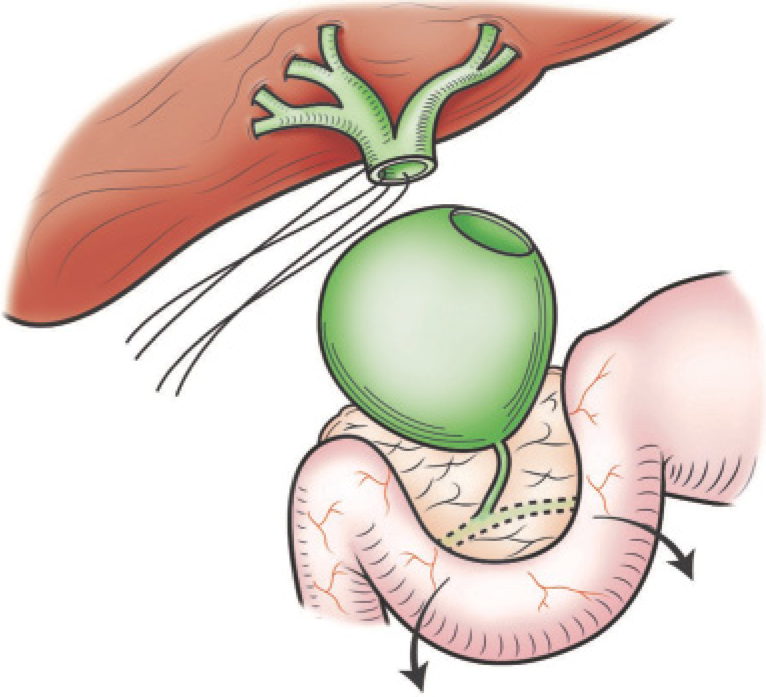
The gallbladder is then separated from the liver bed by cautery and removed in a retrograde fashion down to the cystic duct (**Figure 56.2**). A cholangiogram via the cystic duct or directly into the CBD should always be performed, looking particularly for proximal bile duct variation and degree of intrahepatic dilatation and/or stenosis, and dilatation or obstruction of the common channel. Identification of the cystic artery leads to the right hepatic artery, which is at risk either in front of or behind the neck of the bile duct.



56.2

The overlying peritoneum should be divided anteriorly and laterally, to get onto the wall of the bile duct. Use precise bipolar diathermy to keep this dissection relatively bloodless and stick to this plane (**Figure 56.3a**). The neck of the CHD is proximally delineated up to the bifurcation, a sling is passed and it is divided under vision. Mobilization of the rest of common duct down to the duodenum and pancreas can be done, avoiding the common hepatic artery medially and a potential aberrant right hepatic artery. The portal vein is at risk at the back of the duct. Sometimes, repeated infections and scarring make removal of the wall extremely hazardous. John Lilly's technique might be useful here and refers to leaving the back wall intact but stripping the mucosa off.

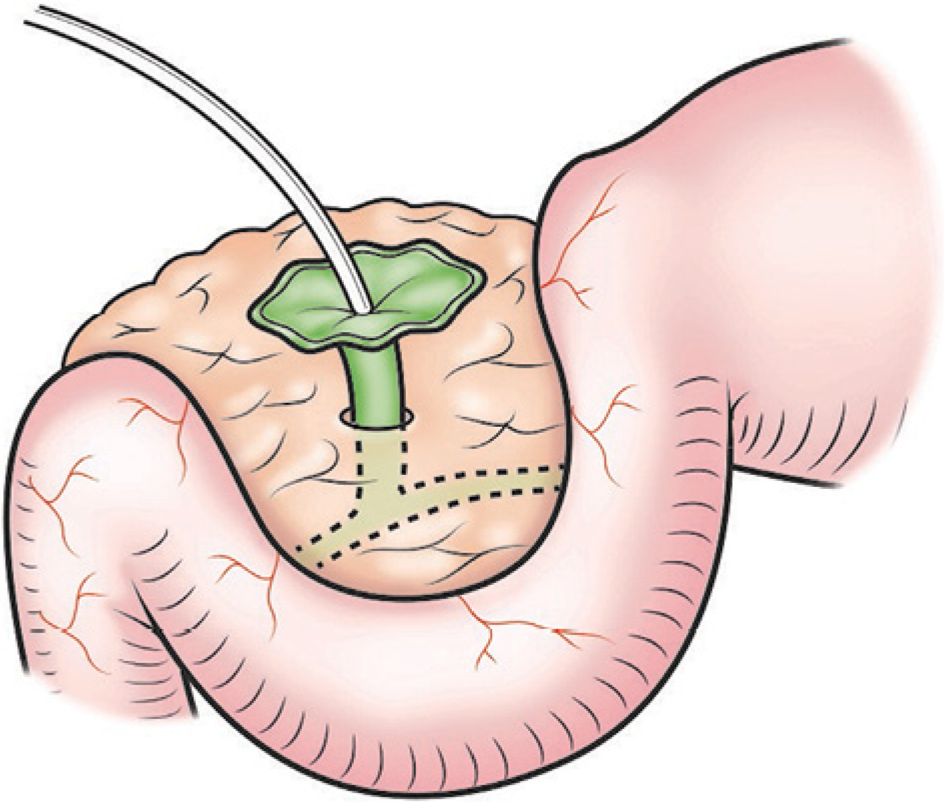

(a)


(b)

56.3a,b

The head of the pancreas is dissected using bipolar diathermy, as the duct narrows down. This can be abrupt in the cystic variants (Types 1C and 4C), leaving a small, thin-walled structure down to the pancreatic duct (**Figure 56.3b**).

The distal duct is divided, leaving a small cuff. A probe or lacrimal duct dilator should be inserted through the ampulla to ensure that there is no stenosis (**Figure 56.4**). Then a small irrigating umbilical vein catheter or, in the older child or adolescent, a flexible choledochoscope can be used to ascertain that the residual parts are stone- and debris-free. The stump is oversewn using an absorbable PDS□ suture.

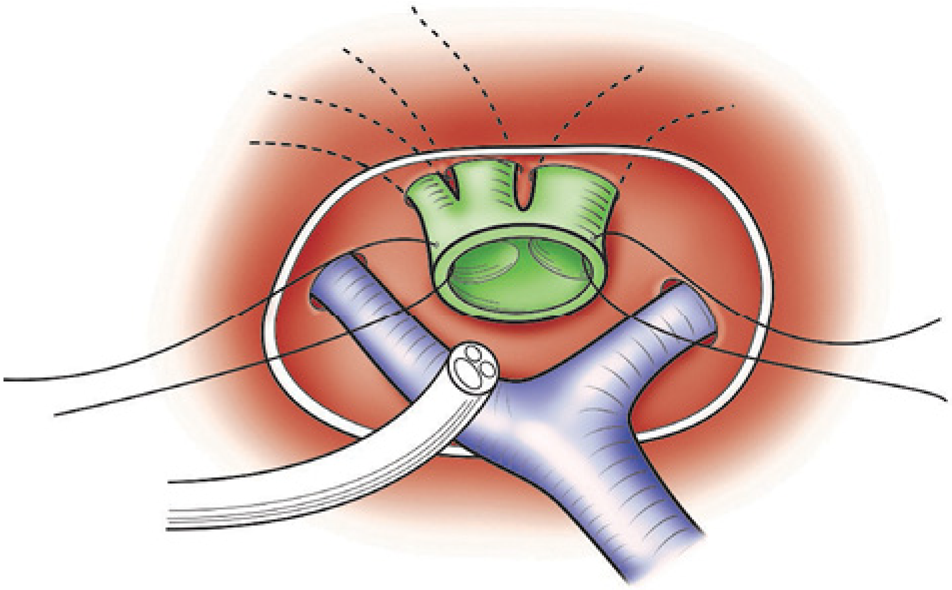


56.4

The proximal pancreatic duct is at risk medially and can be damaged. Suspicion can be confirmed by irrigation using methylene blue or dilute

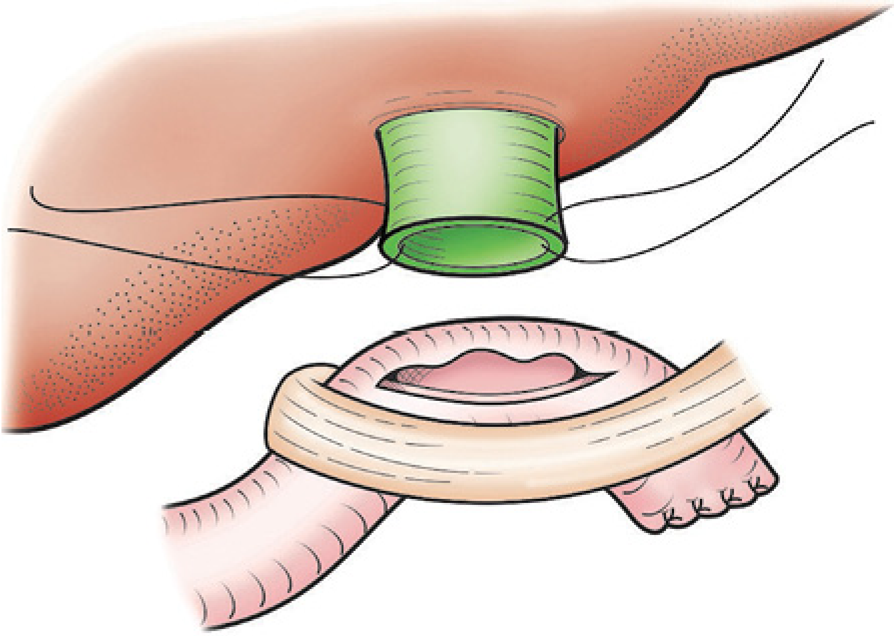
Betadine□. Any damaged area can be oversewn using a fine absorbable monofilament (e.g. 7/0 PDS□). Actual transaction should, in addition, probably be stented using a fine "feeding" tube brought out through the stump of the distal bile duct.

A flexible choledochoscope should be passed proximally into any dilated intrahepatic ducts looking for stones, debris, and any stenotic segments (**Figure 56.5**). Bile from all intrahepatic segments needs to be free to drain and this is by far the best time to correct any areas of stenosis.



56.5

A Roux loop is prepared, starting with mobilizing and then dividing the proximal jejunum 5–10 cm from the duodenojejunal flexure using a GIA linear stapler. About 40 cm of stretched antimesenteric border should be measured and a suitable end-to-side or stapled enteroenterostomy created. The stapled free end is passed through a mesocolic window on the right of the middle colic artery to lie comfortably without tension adjacent to the transected bile duct (**Figure 56.6**).



56.6

The end-to-side hepaticojejunostomy should be done below the bifurcation in most cases. Typically, the back row of sutures is placed first, untied and then "parachuted" down to the porta when complete. Interrupted absorbable monofilament (e.g. PDS®, Monocryl®, Maxon®) sutures are appropriate for children (6/0 or 7/0 for neonates; 4/0 or 5/0 for older children), placed with loupe magnification. The anterior row is then completed and tested for leaks. The mesocolic window should be closed around the Roux loop.

Bespoke surgery

The hepaticojejunostomy may need to be tailored to fit variation in proximal bile duct anatomy (ductoplasty). Typically, an anterior or posterior right hepatic duct may be separate from the main confluence and is then usually stenotic. This needs opening back to a more dilated part and may need a separate opening in the Roux loop. The anastomosis can be done over a small silicone tube to avoid narrowing. The CHD may also be narrow and may require opening anteriorly to spatulate the junction.

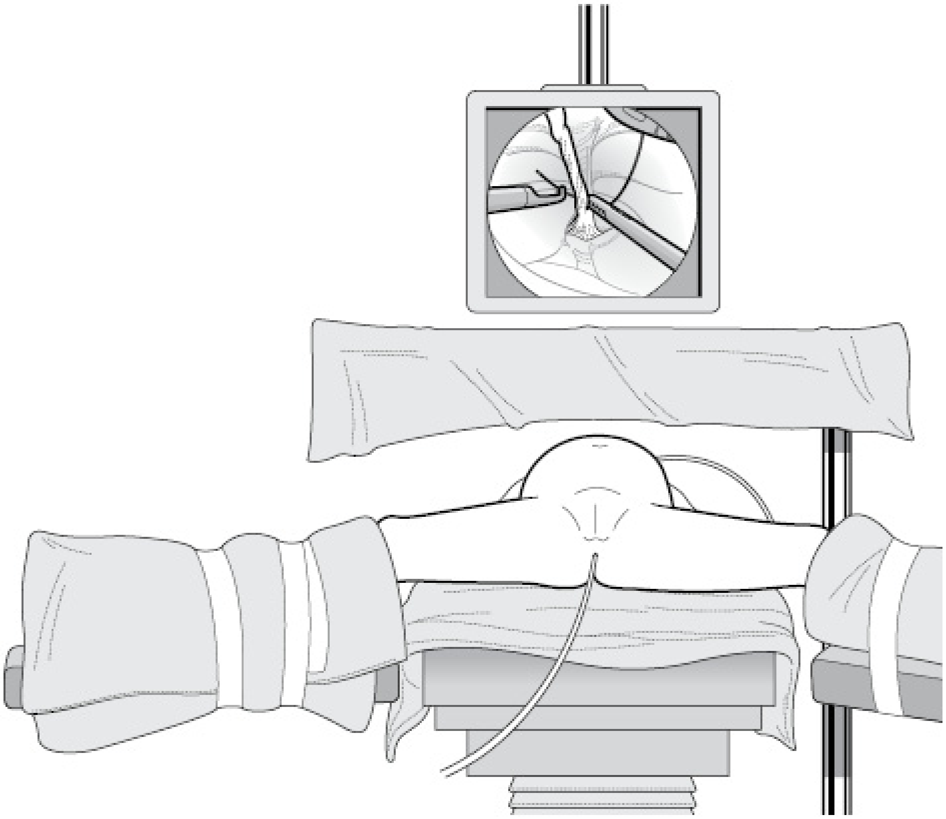
Most surgeons use the antimesenteric border for the top anastomosis, although some favor an end-to-end arrangement. The advantage of the former is that geometrically it is under less tension and the length of the opening can be changed depending on the diameter of the bile duct to which it is to be joined. There is no advantage of introducing any kind of intussusception “valve” in the Roux limb. Postoperative cholangititis is very uncommon in CCM (as compared to biliary atresia).

A transduodenal sphincteroplasty may be warranted if there is a dilated common channel and doubts about natural drainage.

There is a degree of controversy concerning the nature of surgery for patients with Type 4 lesions. In our experience, in children, the intrahepatic dilatation nearly always diminishes postoperatively — often to normal values — providing there is a wide hepaticojejunostomy reducing intrabiliary pressure and all the dilated parts are demonstrably drained. In adults with such pathology, an accompanying resection (typically for left-sided dilatation) may be reasonable.

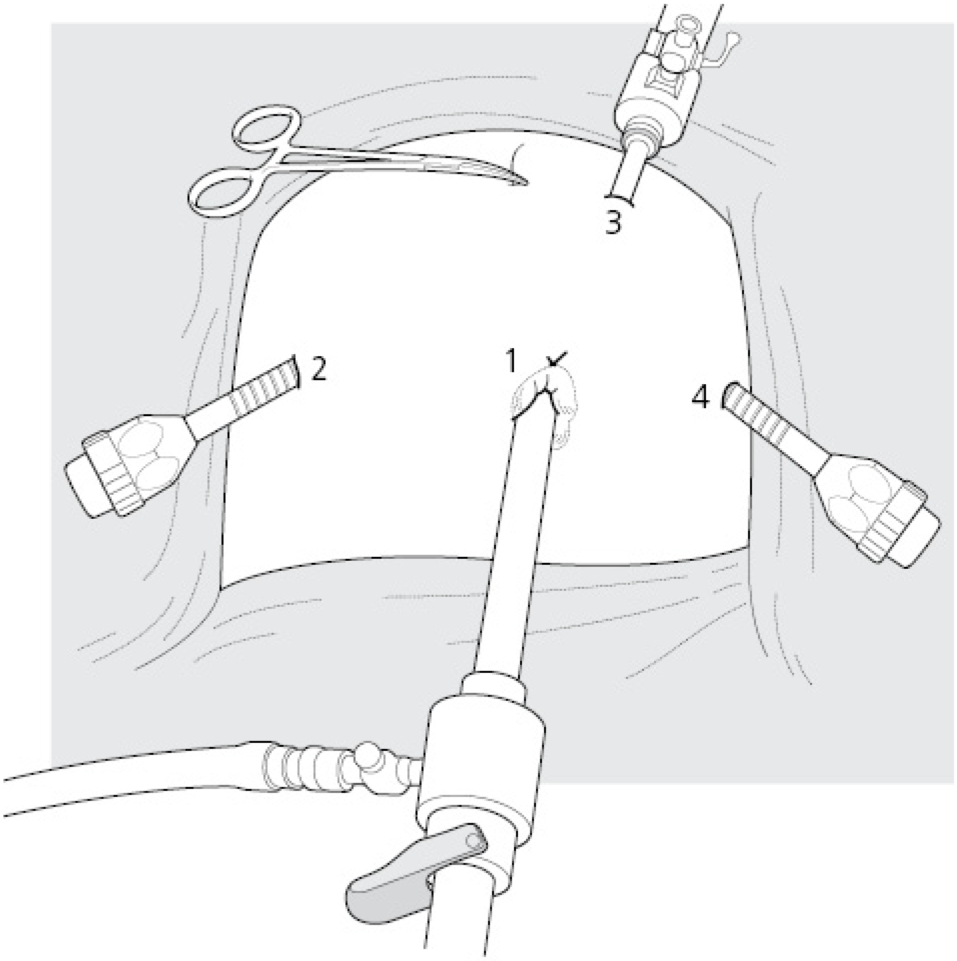
LAPAROSCOPIC SURGERY

A nasogastric tube, rectal tube, and urinary catheter are used to decompress the stomach, the colon, and bladder, respectively. The patient is placed in a 30-degree head-up supine position. The surgeon stands at the lower end of the operating table between the patient’s legs (**Figure 56.7**).



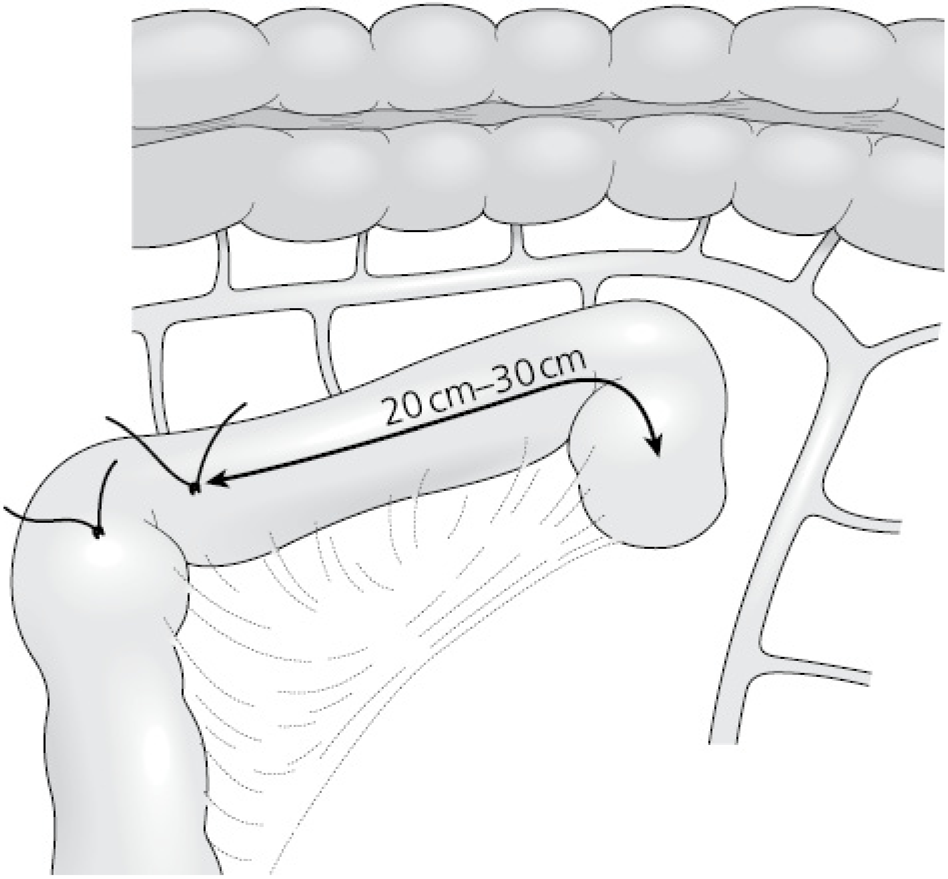
56.7

A 10 mm trocar is inserted through the umbilicus for the telescope. Three additional 5 mm or 3 mm trocars are placed for instruments: One at the right flank, one at the left flank, and the final one in the left hypochondrium (**Figure 56.8**).



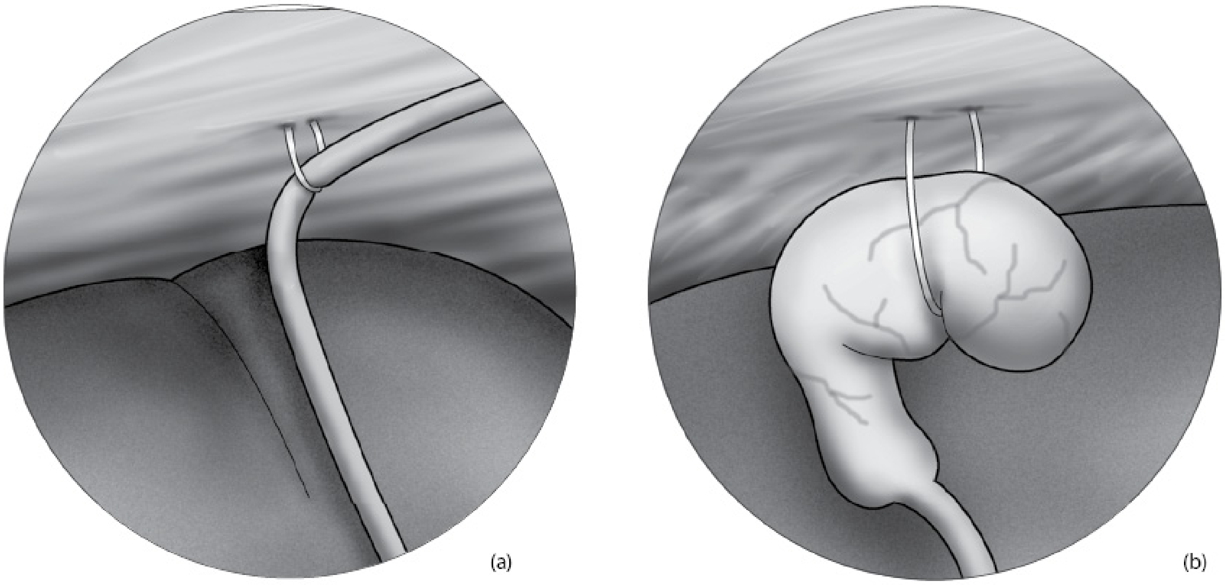
56.8

Carbon dioxide pneumoperitoneum is maintained at a pressure of 8–12 mmHg. The ligament of Treitz is identified by laparoscopy. A 5/0 silk stay suture is placed 20–30 cm distal to the ligament of Treitz. A second 5/0 PDS suture is placed 2.0 cm below the first sutures to mark the jejunal limb, which will be anastomosed to the hepatic duct (**Figure 56.9**). The jejunal segment with two sutures is grasped with an intestinal grasper. The transumbilical vertical incision is extended 1.0 cm above the umbilicus. The jejunum is brought out, and the jejunostomy is carried out extracorporeally. The jejunum is then reintroduced into the abdominal cavity. The extended incision is closed. The laparoscopic instruments are repositioned.



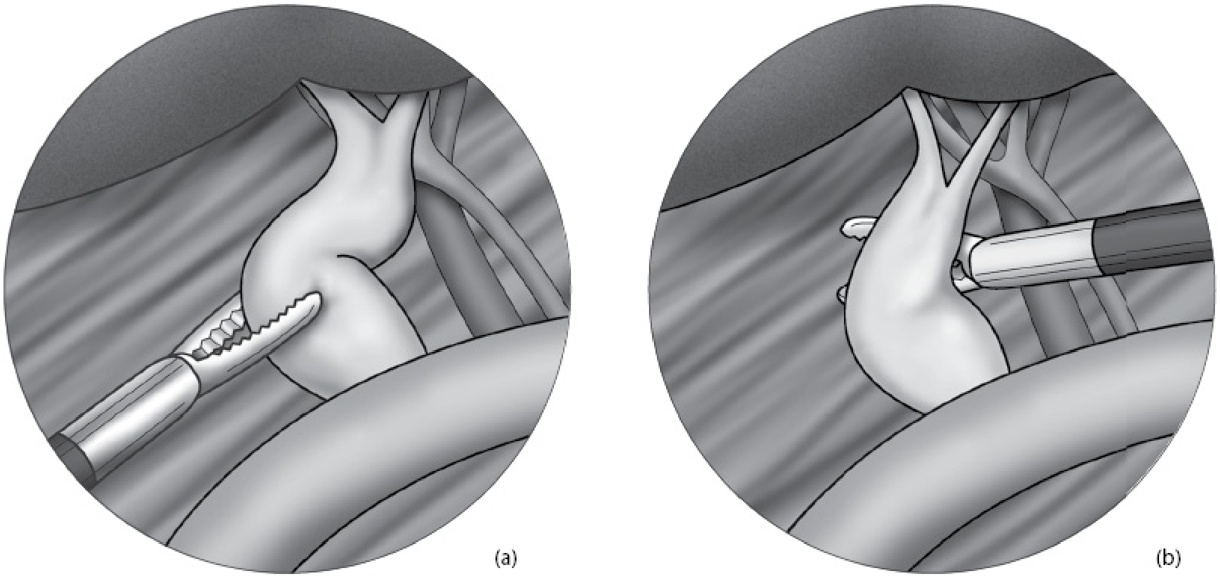
56.9

The liver is secured to the abdominal wall by a stay suture placed at the round ligaments (**Figure 56.10a**). The cystic artery is identified, clipped, and divided. The cystic duct is also isolated, clipped, and divided. A second traction suture is placed at the distal cystic duct to elevate the liver and splay out the liver hilum (**Figure 56.10b**).



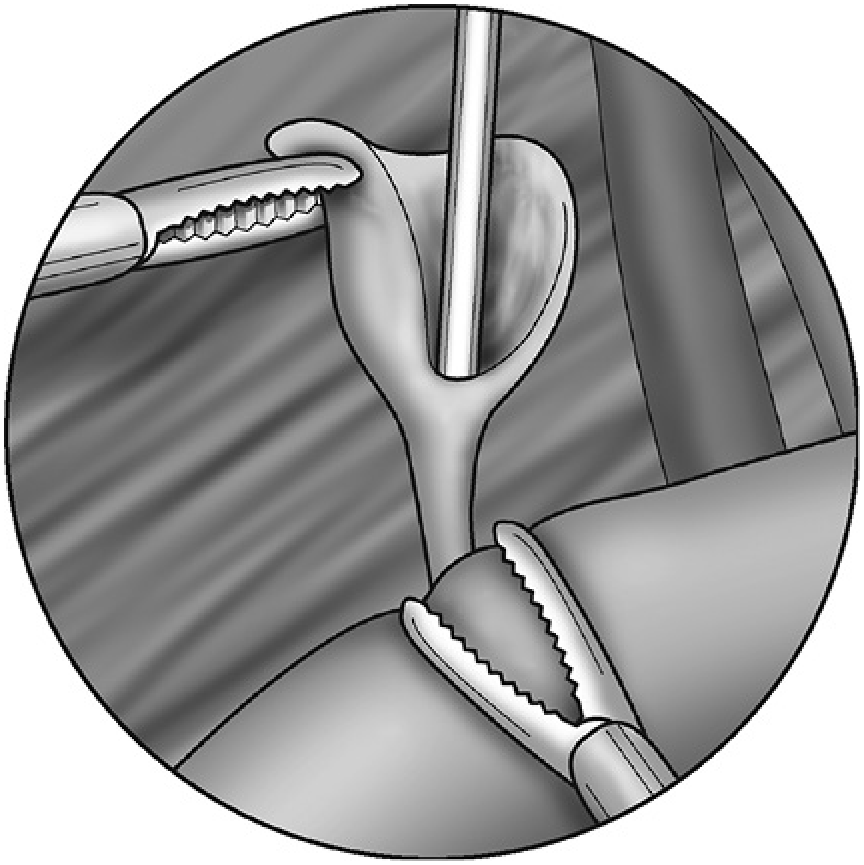
56.10a,b

The duodenum is retracted downward using a dissector through the fourth trocar site (**Figure 56.11a**). The mid-portion of the cyst is dissected circumferentially. Separation of the cyst from the hepatic artery and portal vein is carried out meticulously until a dissector can be passed through the space between the posterior wall of the cyst and portal vein proceeding from left to right (**Figure 56.11b**).



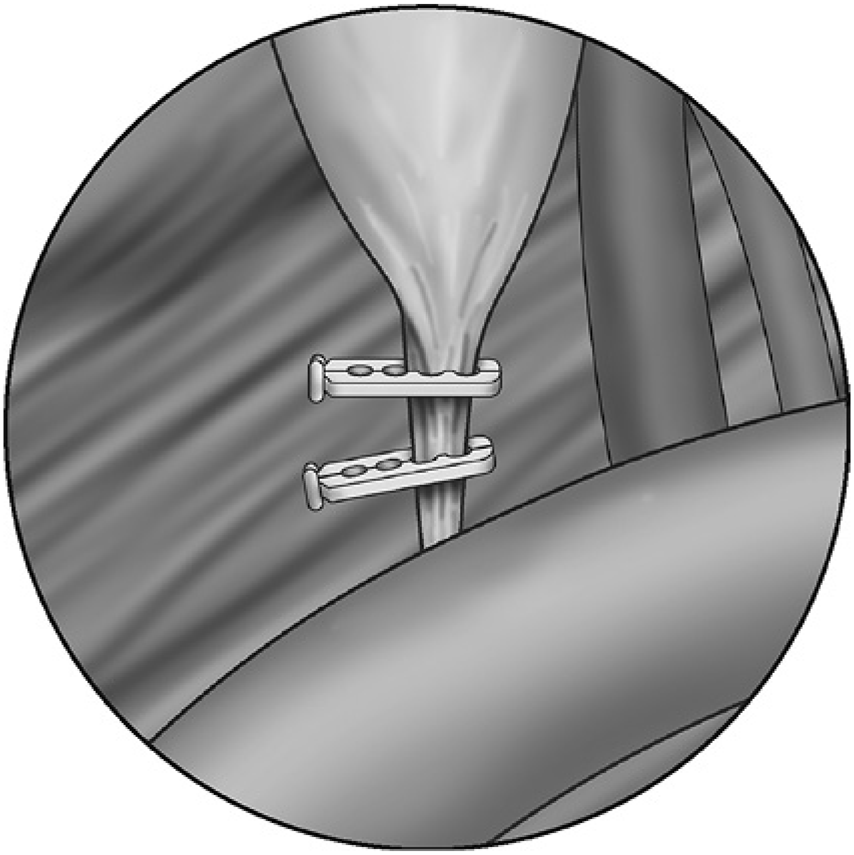
56.11a,b

The cyst is then divided at this site. The lower part of the cyst is detached from the pancreatic tissue down to the common biliary–pancreatic duct using a 3 mm dissector for cautery and dissector for cautery and dissection (**Figure 56.12**). The distal part of the cyst is opened longitudinally. The interior of the cyst is inspected to identify the orifice of the common channel. Irrigation is performed with normal saline via a catheter which is inserted into the common channel then passed down to the duodenum.



56.12

The inspection and irrigation may be performed through a pediatric cystoscope if the the common channel is wide enough. The distal choledochal cyst is clipped and divided at the level of the orifice of the common channel (**Figure 56.13**).



56.13

The upper part of the cyst is now dissected up to the CHD and divided. The choledochal cyst is initially divided at the level of the cystic ducts. The definitive division is performed after identifyng the orifice of the CHD. The CHD is irrigated with normal saline to wash out biliary debris and calculi. Irrigation with normal saline through a small catheter inserted into the right and then into the left hepatic duct is performed to wash out the protein plugs or calculi until the fluid from those ducts is clear.

With a large cyst, the dissection is started at the middle portion, proceeding distally. The distal portion of the choledochal cyst is separated from the portal vein. The distal CBD is divided above the biliary–pancreatic duct. Irrigation with normal saline in the remnant is performed to wash out debris and calculi. The distal CBD is then clipped and transected at the level of the common channel orifice.

When the cyst is intensely inflamed and extensive pericystic adhesion is present, the dissection of cystic wall from the portal vein is carried out carefully while viewing the cyst internally and externally. After dividing the mid-portion of the cyst, the upper and lower parts of the cyst are removed.

The Roux limb is passed through a window in the transverse mesocolon to the porta hepatic. The jejunum is opened longitudinally on the antimesenteric border a few millimeters from the end of the Roux loop. Hepaticojejunostomy is fashioned using two running sutures of 5/0 PDS. (Interrupted sutures are used when the diameter of the commen hepatic duct is <1 cm.) Sutures are inserted from the left to right with 3 mm instruments, and a 3 mm needle driver is used to suture. Ductopasty is performed by opening the CHD and incising the left hepatic duct if it is too small.

Mesenteric defects in the transverse mesocolon and small bowel mesentery are closed with sutures.

The gallbladder is detached from its bed and surrounding tissues. Different parts of the cyst and gallbladder are removed through the umbilicus. The operative field is washed with warm saline and a subhepatic drain is inserted.

Laparoendoscopic single-site surgery with conventional instruments

A Z-shaped incision is made on the umbilicus and the skin is detached from the fascia. A 5 mm trocar is inserted in the middle for the conventional 30-degree laparoscope, and two other trocars (one 5 mm and one 3 mm) are introduced in the left and the right for instruments. A transabdominal suspending suture is made to lift the hepatic round ligament to the abdominal wall. A second suspending suture is performed to lift the gallbladder to the abdominal wall on the right. A third suspending suture is made to lift the anterior wall of the choledochus to the abdominal wall. More suspending sutures might be needed to facilitate dissection of the choledochus distally during the operation. The other parts of the operation are performed as described in the section on “Laparascopic Surgery”.

OUTCOME

Morbidity from this kind of surgery is uncommon (<5%).

COMPLICATIONS

Early complications

Early complications might include bleeding and bile leakage from the anastomosis. The latter may be evident early if a drain has been used or with increasing right upper quadrant pain and tenderness, together with pyrexia, and failure to re-establish feeding if not. Most leaks are minor and technical in origin and they usually respond to drainage of any collection. If a fistula develops, there should be concern over the drainage of the Roux loop. A radio-isotope hepatobiliary iminodiacetic acid (HIDA) scan should show whether the leak is partial or total.

Pancreatic leakage may also occur from duct laceration or leak from the oversewn distal duct. Measurement of amylase levels should confirm its nature. Again, this should be treatable with conservative measures, attention to nutrition, and occasionally a period of parenteral nutrition. Octreotide may be tried. If there is an ongoing leak sometime following surgery, ERCP should be performed to delineate its site.

Late complications

As stated above, cholangitis is very uncommon following bile duct reconstruction. Confirmation of this diagnosis mandates thorough investigation of the Roux loop and proximal anastomosis by US, radio-isotope studies, and possibly percutaneous transhepatic cholangiography (PTC).

Pancreatitis may occur later and the usual cause is related to a recurrence of protein plug debris in the common channel. This should respond to endoscopic clearance ± sphincterotomy.

Persistence of intrahepatic biliary dilatation, and stone formation as a consequence, implies poor drainage. In historical Japanese series, this was estimated to occur in up to 5% of cases, but it is now seen less often.

While the epithelium of a CCM has an undoubted risk of carcinomatous change, this is not really a practical problem during childhood. By contrast, up to 10% of adult series with a mean age of about 40 years have established malignant change at laparotomy. We do not know the precise risk in later life as our children will have had a radical resection of a CCM. Our practice is to encourage appropriate follow-up, ideally by serial ultrasonography, particularly for those with Type 4 lesions.

VIDEO

OPS_8e_video_56.1 Laparoscopic choledochal cyst

FURTHER READING

Friedmacher F, Ford KE, Davenport M. Choledochal malformations: Global research, scientific advances and key controversies. *Pediatr Surg Int* 2019; 35(3): 273–82.

Hill R, Parsons C, Farrant P, et al. Intrahepatic duct dilatation in type 4 choledochal malformation: Pressure-related, postoperative resolution. *J Pediatr Surg* 2011; 46: 299–303.

Kronfli R, Davenport M. Insights into the pathophysiology and classification of Type 4 choledochal malformation. *J Pediatr Surg* 2020; S0022-3468(20)30330-4. doi: 10.1016/j.jpedsurg.2020.05.017.

Liem NT, Pham HD, Dung le A, et al. Early and intermediate outcomes of laparoscopic surgery for choledochal cysts with 400 patients. *J Laparoendosc Adv Surg Tech A* 2012; 22: 599–603.

Son TN, Liem NT, Hoan VX. Transumbilical laparoendoscopic single-site surgery with conventional instruments for choledochal cyst in children: Early results of 86 cases. *J Laparoendosc Adv Surg Tech A* 2014; 24: 907–10.

Turowski C, Knisely AS, Davenport M. Role of pressure and pancreatic reflux in the aetiology of choledochal malformation. *Br J Surg* 2011; 98: 1319–26.

57

Splenectomy

MARCUS D. JARBOE AND STEVEN W. BRUCH

PRINCIPLES AND JUSTIFICATION

Children require splenectomies mainly to treat hematologic disorders, remove masses, and control bleeding after trauma. The open and laparoscopic approaches each have their place depending on the clinical scenario surrounding the splenectomy. The laparoscopic approach results in reduced pain, shorter hospital stay, and improved cosmesis, making it the preferred approach. However, certain situations, such as trauma, benefit from the open approach. When considering splenectomy, the potential benefits must outweigh the postoperative risks of overwhelming post-splenectomy sepsis, thrombosis, and pulmonary hypertension. The risk of complete splenectomy has led to the development of the partial splenectomy, with a goal of reducing the burden of hematologic disorders or removing the damaged or abnormal spleen while maintaining enough function to avoid postoperative risks, especially the risk of overwhelming post-splenectomy sepsis.

INDICATIONS

- Hematologic disorders:
  - Hereditary spherocytosis
  - Sickle cell anemia
  - Alpha and beta thalassemia

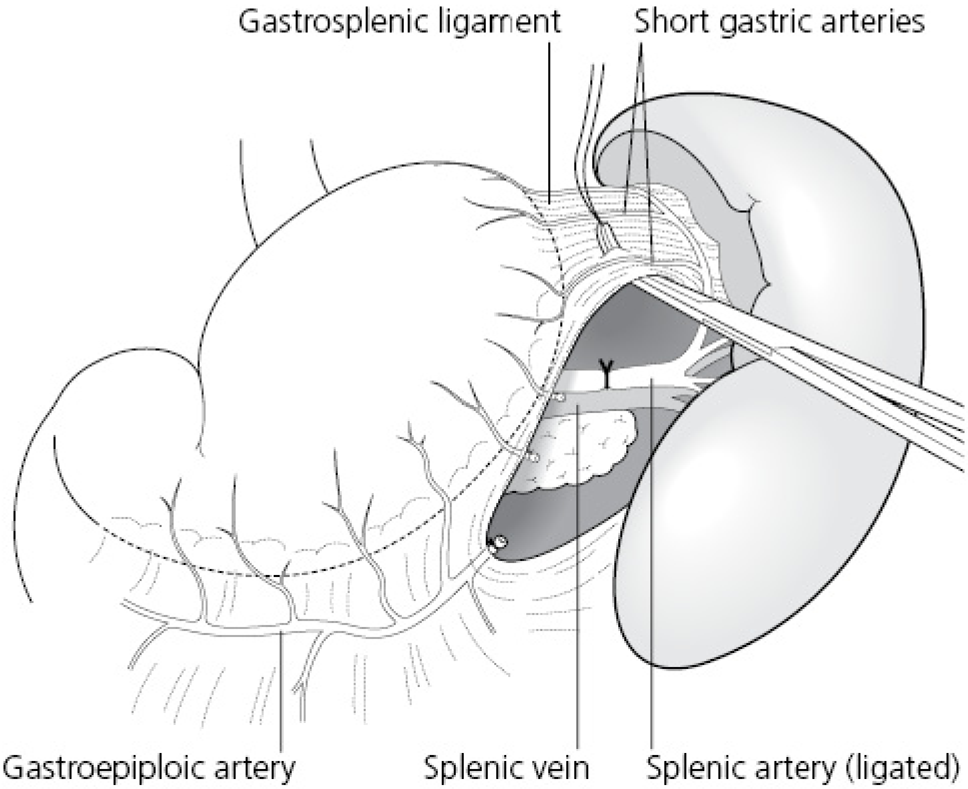
  - Idiopathic thrombocytopenic purpura
  - Gaucher’s disease
- Masses or cystic lesions
- Trauma

PREOPERATIVE PREPARATION

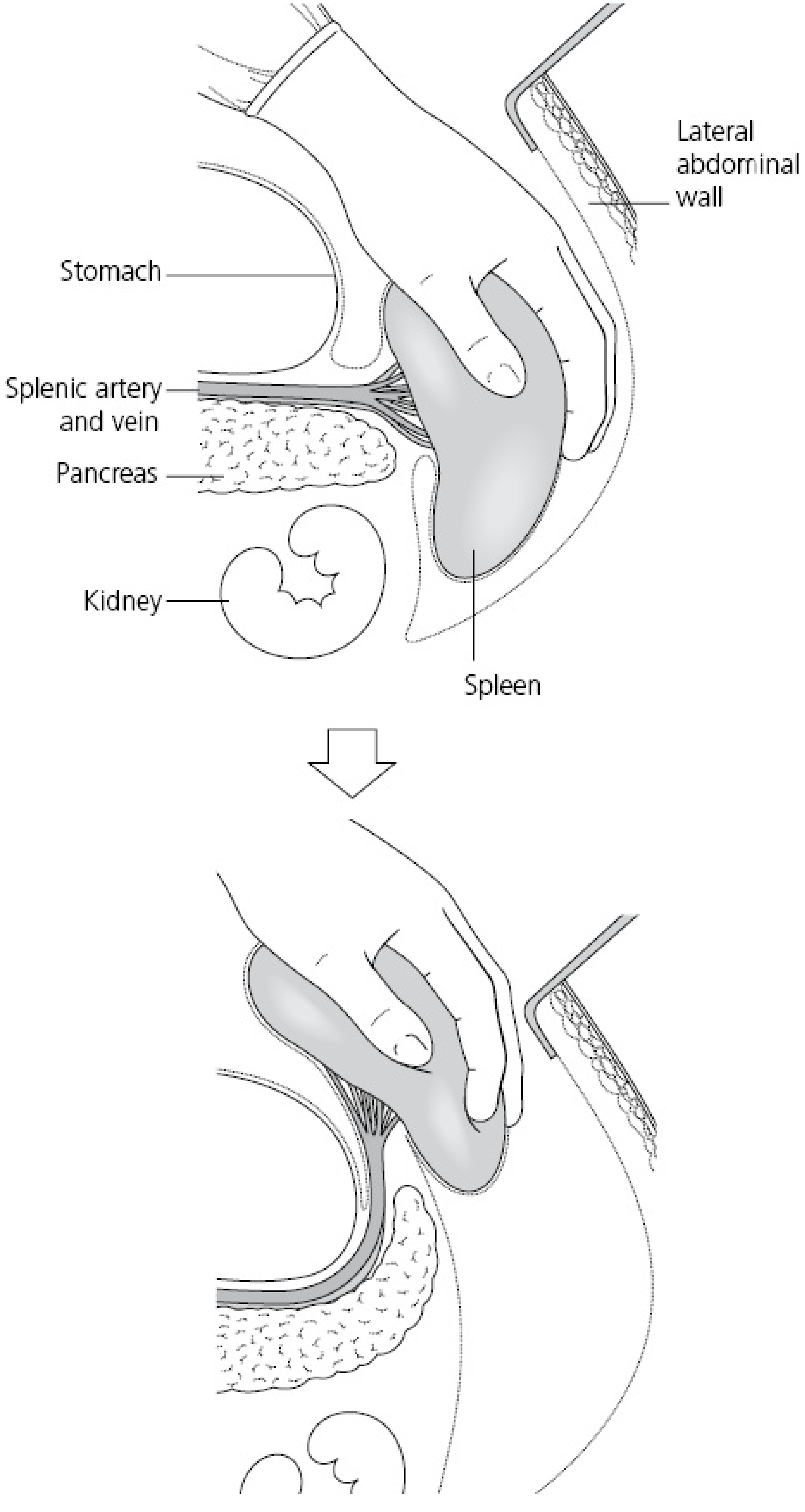
To prevent infection with encapsulated bacteria after a splenectomy, patients receive immunizations to *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Haemophilus influenzae* at least 2 weeks prior to the date of operation. Trauma patients requiring splenectomy should receive these immunizations after recovering from the splenectomy. When removing the spleen for a hematologic disorder, a right upper quadrant ultrasound noting gallstones prompts a cholecystectomy at the time of splenectomy. Sickle cell anemia patients often require admission the day prior to the splenectomy for hydration and possible transfusion to attempt to avoid a sickle cell crisis in the immediate postoperative period.

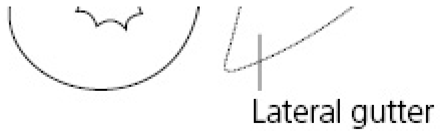
OPERATIONS

A splenectomy requires separation of the spleen from its blood supply, and from its ligamentous attachments. The splenic artery and vein provide the majority of the blood supply with the short gastric vessels supplying a portion to the upper pole (**Figure 57.1**). The splenic artery and vein divide prior to entering the splenic hilum, allowing division of the main pedicles or the individual branches entering the splenic capsule. Several methods exist to control these vessels including ligatures, stapling devices, and thermal energy devices (LigaSure® [Valleylab, Boulder, CO, USA], Harmonic scalpel® [Ethicon Inc., Cincinnati, OH, USA]). The proximity of the tail of the pancreas to the splenic hilum dictates meticulous dissection of the vessels to avoid injury to the pancreatic parenchyma. Ligaments attach the spleen to the lateral abdominal wall, colon, kidney, stomach, and diaphragm (**Figure 57.2**). These require division to expose the spleen in open operations but are mostly left intact until the end of laparoscopic operations to suspend the spleen to gain adequate exposure for vascular division.


Gastrosplenic ligament
Short gastric arteries
Gastroepiploic artery
Splenic vein
Splenic artery (ligated)

57.1

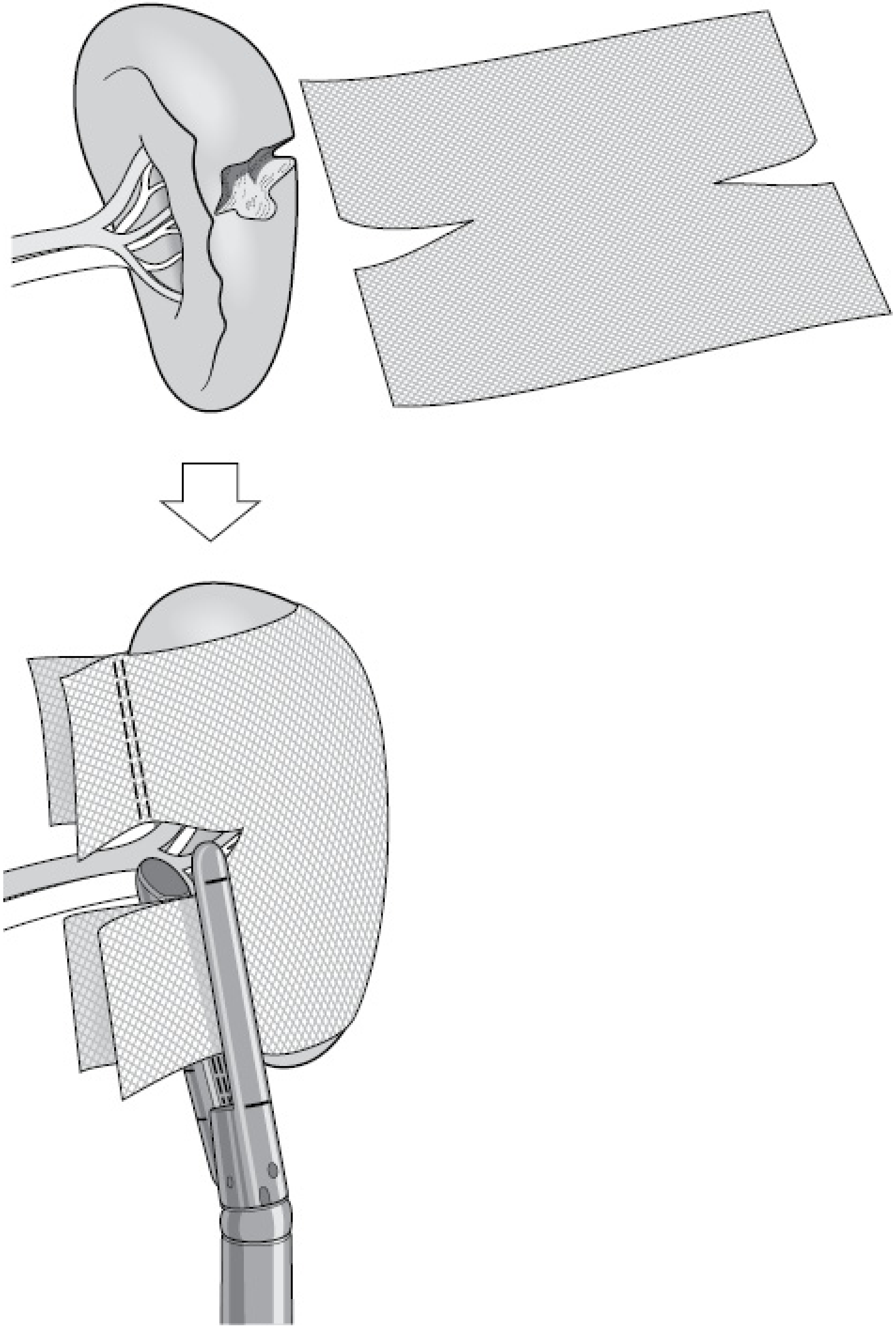

Lateral abdominal wall
Stomach
Splenic artery and vein
Pancreas
Kidney
Spleen



57.2

Splenic trauma

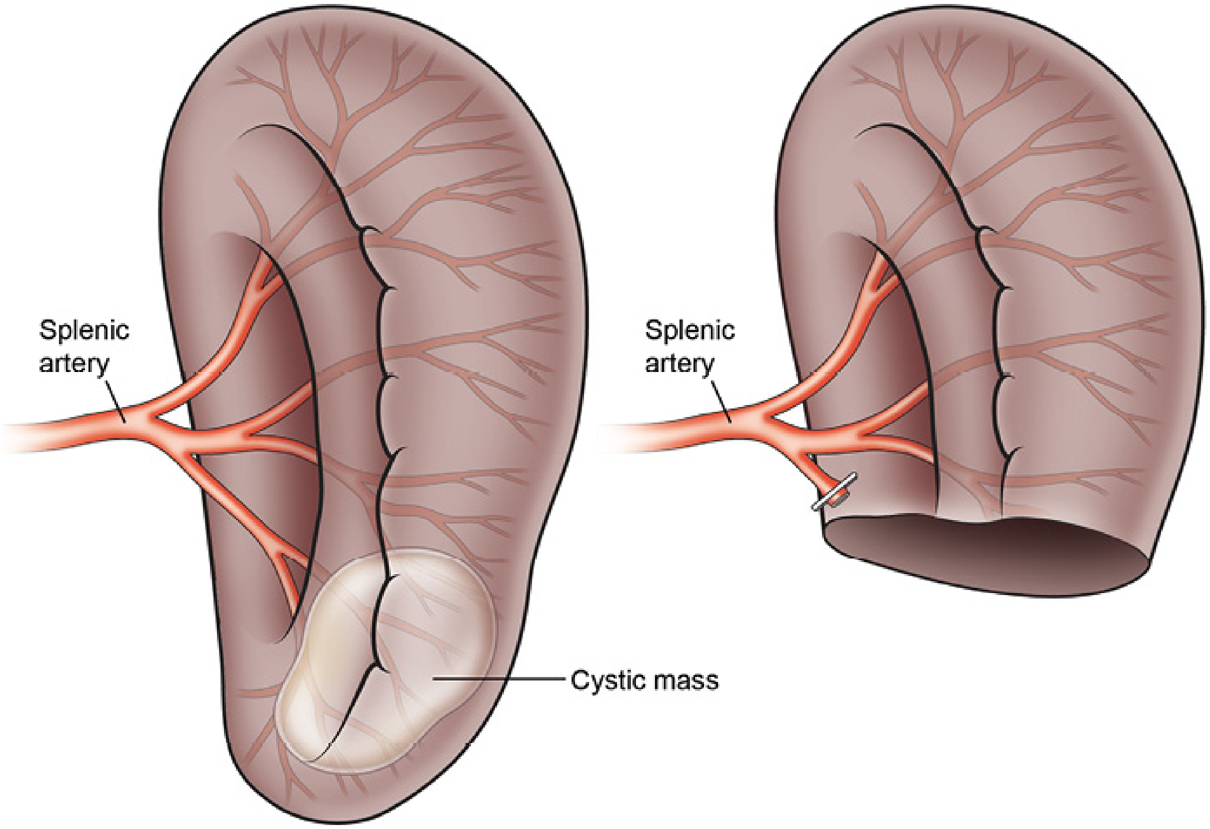
An open midline incision allows inspection of the remainder of the abdomen while providing excellent exposure for splenectomy. To gain vascular exposure, rapid division of the ligamentous attachments allows mobilization of the spleen out of the abdomen. Left upper quadrant packing provides time to assess the injury and decide if a total splenectomy, partial splenectomy, or splenorrhaphy optimally treats the injury. In addition to the anatomy of the injury, the stability of the patient determines the eventual treatment. With a hemodynamically stable patient, a partial splenectomy or splenorrhaphy retains the immunologic function of the spleen. A splenorrhaphy attempts to compress the spleen using an absorbable mesh. Two slits in a rectangular piece of mesh allow the mesh to wrap around the spleen with room for the vascular pedicle to enter. A stapling device or running suture then closes the mesh around the spleen both superior and inferior to the splenic vessels, allowing compression of the splenic tissue, while maintaining vascular inflow and return by the splenic artery and vein (**Figure 57.3**).



57.3

Splenic cysts or masses

Depending on its position, a partial splenectomy allows removal of the cystic or solid lesion while maintaining the remainder of the spleen. Often aspiration or unroofing of a splenic cyst results in recurrence making removal the preferred treatment. Cystic or mass lesions in the center of the spleen often require a total splenectomy, while polar lesions allow the option of a partial splenectomy (**Figure 57.4**). The size of the lesion and preference of the surgeon determines the approach, open or laparoscopic. The open operation involves either an upper midline incision or a left upper quadrant incision with the patient supine, or a left upper quadrant incision with the patient in the right lateral decubitus position (which is the authors' preferred exposure for an open elective splenectomy).

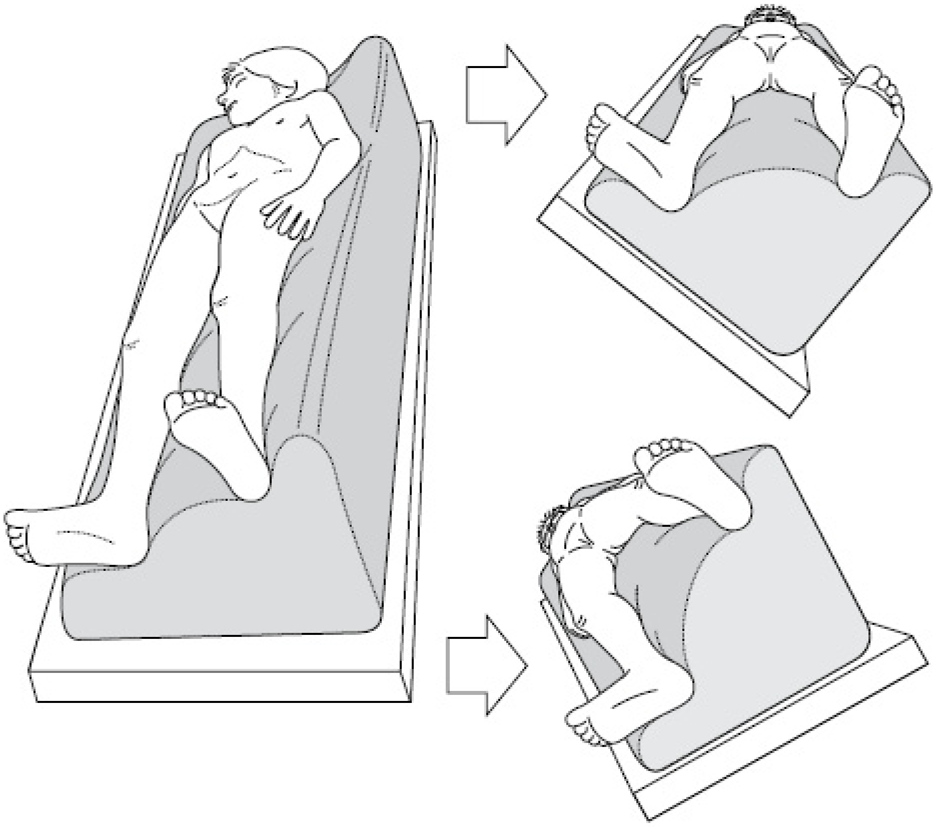


57.4

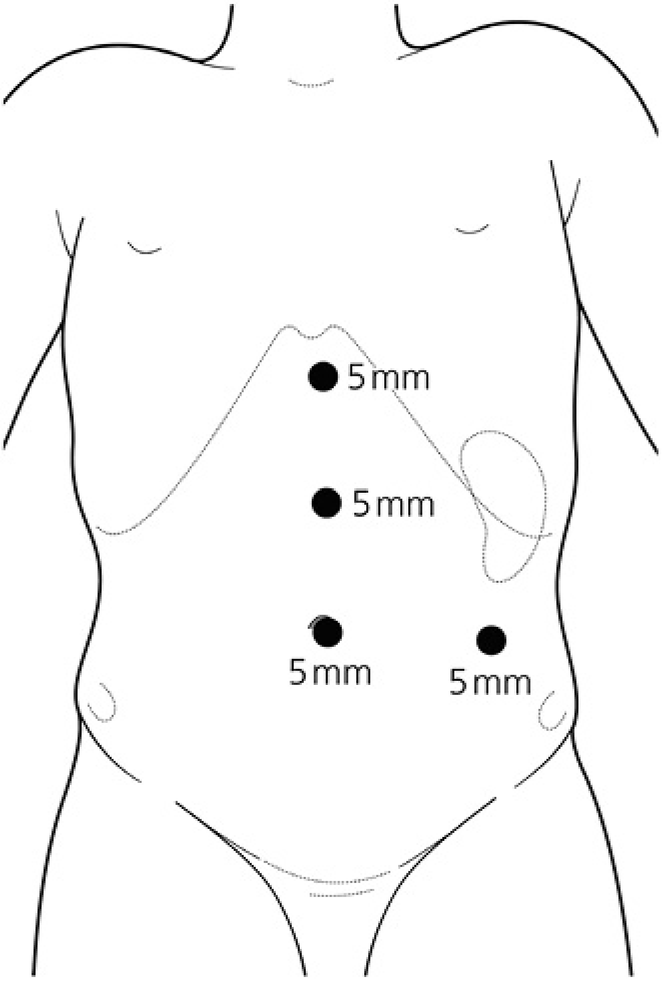
Hematologic disease

The spleens that require removal for hematologic reasons often result from splenic sequestration resulting in large spleens with the tips below the left costal margins. Most surgeons now utilize the laparoscopic approach to remove these spleens. The introduction of the partial splenectomy attempts to provide adequate treatment of the hematologic problem, reducing transfusion requirements, while maintaining the immunologic function of the spleen, reducing the incidence of overwhelming post-splenectomy sepsis.

For laparoscopic splenectomy, patients need to be positioned close to supine for trocar placement and removal but tilted with the left side up for the dissection (**Figure 57.5**). Use of a bean bag allows the table tilt to achieve these positions. Optimal trocar sites depend on the size of the spleen and the size of the patient. In the pediatric population, an umbilical trocar for the camera, a subxiphoid trocar for retraction laterally, and two dissecting trocars (one halfway between the xiphoid and the umbilicus and the other in line with the splenic hilum just inferior to the lower pole of the spleen) provide excellent ergonomics (**Figure 57.6**). In larger children, one should consider placing the four trocars along but just inferior to the costal margin.



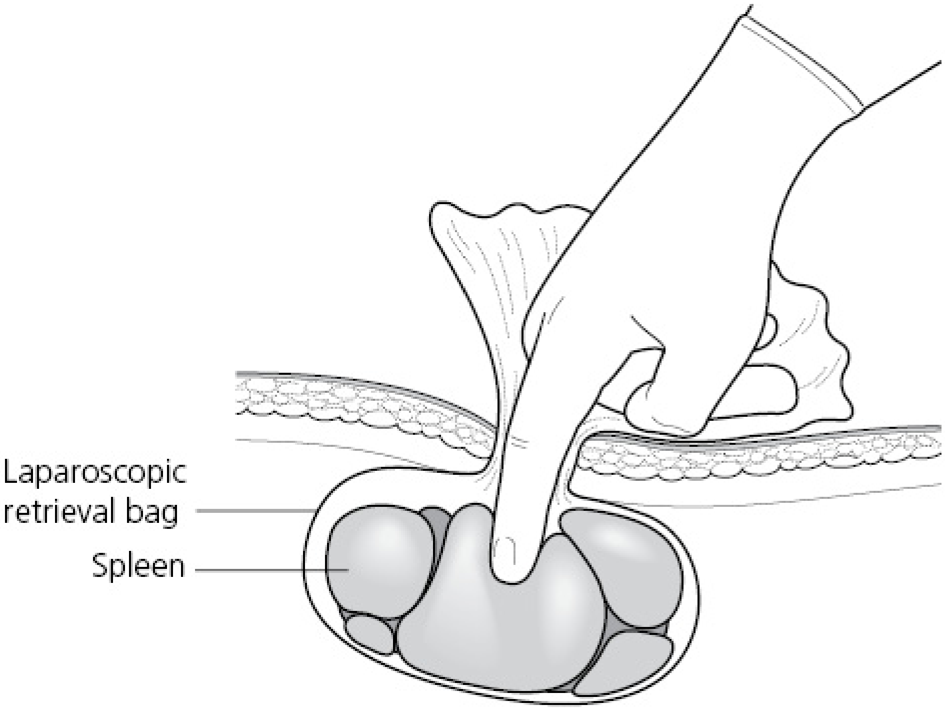
57.5



57.6

Laparoscopic dissection

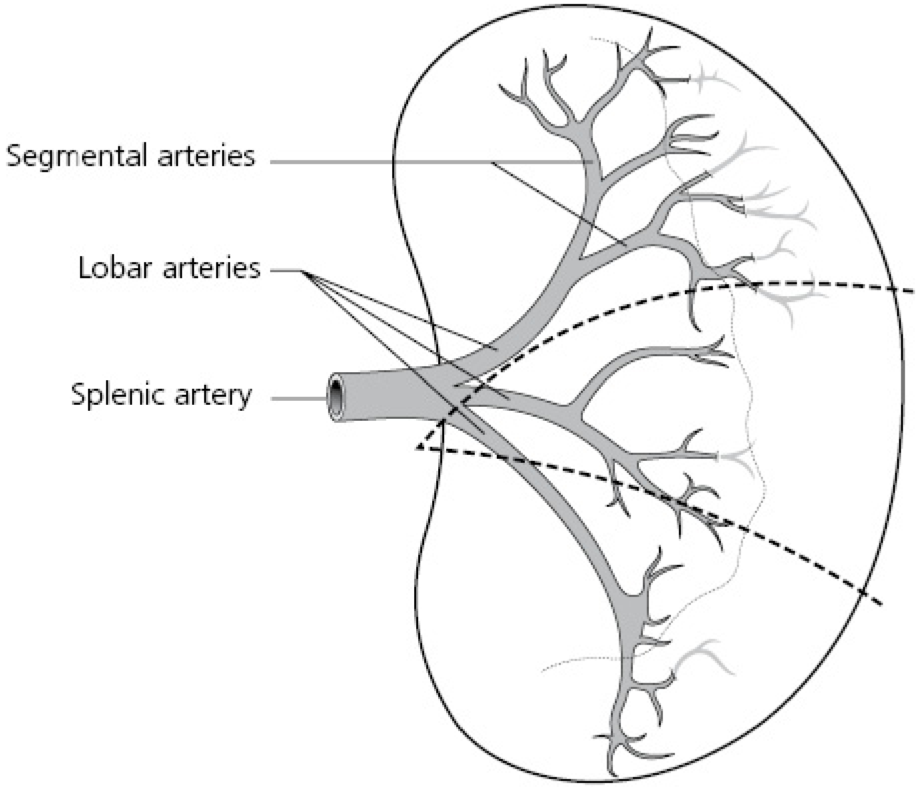
In contradistinction to the open operation, the ligamentous attachments to the spleen remain intact until the end of the procedure, allowing the spleen to hang from the lateral abdominal wall while in right lateral decubitus position exposing the vascular pedicle during the dissection. Opening the peritoneum overlying the hilum of the spleen allows excellent visualization of the splenic artery and vein and its bifurcation pattern entering the spleen. Two options exist to control the vessels. Dissection and division of the main trunk of the splenic artery then vein using a vascular stapler or energy device (LigaSure™ [Valleylab, Boulder, CO, USA]; or Harmonic scalpel™ [Ethicon Inc., Cincinnati, OH, USA]) depending on the size of the vessels. This allows early control of the vessels, but risks injury to the tail of the pancreas. The other option involves dividing individual branches of the splenic artery and vein at the level of the splenic capsule using an energy device. This dissection occurs further away from the tail of the pancreas but requires delicate technique to avoid bleeding from the many vessels. The short gastric vessels then require division. This devascularizes the spleen. Division of the ligaments attaching the spleen to the lateral side wall while leaving a small attachment from the superior pole of the spleen to the diaphragm stabilizes the spleen, allowing easy placement of the spleen into a laparoscopic retrieval bag (e.g. Endo Catch™; Covidien, Mansfield, MA, USA). Division of the last attachment of the spleen to the diaphragm allows the spleen to drop into the bag. The left lower quadrant trocar incision requires enlargement to accept the 15 mm bag. The bag then emerges from the abdomen, and morsilization of the spleen allows complete extraction without intra-abdominal spill (**Figure 57.7**).

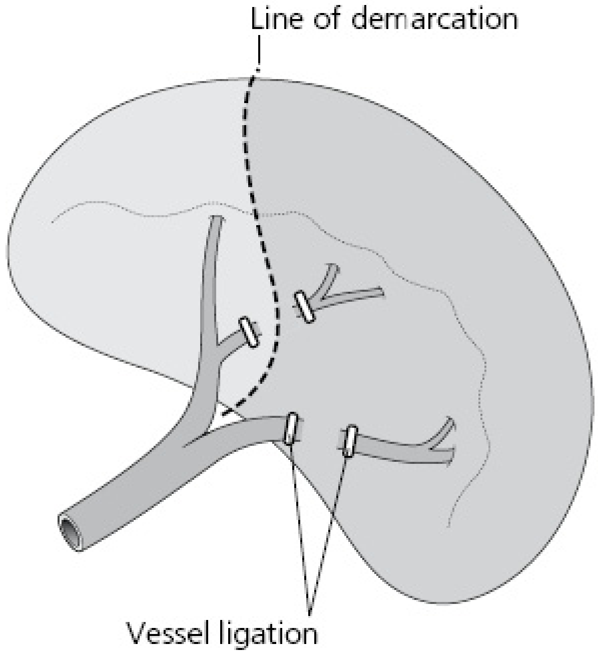


57.7

Laparoscopic partial splenectomy

Partial splenectomy attempts to retain 10–15% of the splenic volume on its vascular pedicle to help prevent postoperative sequelae of a total splenectomy. The operation mimics a total laparoscopic splenectomy with the same patient positioning. Inspection of the vascular supply allows the surgeon to determine whether the superior or the inferior pole appears preferable to leave intact. Most often, the superior pole remains due to some extra blood supply offered by the short gastric vessels, and the more robust ligamentous attachments to the diaphragm, which minimize the risk of the splenic remnant twisting postoperatively. The vessels require dissection and division at the level of the splenic capsule. This allows protection of the last branch of the splenic artery and vein to the upper or lower pole which results in the acceptable size of splenic remnant. The spleen will demarcate showing the extent of the devascularization. When the appropriate amount remains with good blood supply (10–15%), the lateral ligaments up to the level of demarcation require division (**Figure 57.8**). The ligaments attaching the vascularized splenic remnant remain to stabilize the remnant, preventing torsion. In order to minimize blood loss during the division of the spleen, the Harmonic scalpel□ (Ethicon Inc., Cincinnati, OH, USA) divides the parenchyma, staying just on the devascularized side of the spleen. The laparoscopic argon beam coagulator then controls the remaining bleeding from the cut surface. The laparoscopic retrieval bag allows removal of the devascularized spleen through the left lower quadrant incision.


Segmental arteries
Lobar arteries
Splenic artery


Line of demarcation
Vessel ligation

57.8

FURTHER READING

Costi R, Ruiz CC, Romboli A, et al. Partial splenectomy: Who, when, how. A systematic review of the 2130 published cases. *J Pediatr Surg* 2019; 54(8): 1527–38.
Englum BR, Rothman J, Leonard S, et al. Hematologic outcomes after total splenectomy and partial splenectomy for congenital hemolytic anemia. *J Pediatr Surg* 2016; 51: 122–7.
Geiger JD, Dinh VV, Teitelbaum DH, et al. The lateral approach for open splenectomy. *J Pediatr Surg* 1998; 33: 1153–7.
Hirshberg A, Mattox KL. The 'take-outable' solid organs. In: Hirshberg A, Mattox KL (eds). *Top Knife: The Art and Craft of Trauma Surgery*. Harley: tfm Publishing Ltd; 2005: pp. 100–14.
Lobe TE, Presbury GJ, Smith BM, et al. Laparoscopic splenectomy. *Pediatr Ann* 1993; 22: 671–4.
Madenci AL, Armstrong LB, Kwon NK, et al. Incidence and risk factors for sepsis after childhood splenectomy. *J Pediatr Surg* 2019; 54(7): 1445–8.
Reinberg O. Partial splenectomies by laparoscopy in children. In: Bax K, Georgeson KE, Rothenberg SS, et al. (eds). *Endoscopic Surgery in Infants and Children*. Heidelberg: Springer; 2008: pp. 455–61.
Rescorla FJ. Splenectomy. In: Bax K, Georgeson KE, Rothenberg SS, et al. (eds). *Endoscopic Surgery in Infants and Children*. Heidelberg: Springer; 2008: pp. 449–54.

PART V

Tumors

58

Liver resection

REBECKA L. MEYERS, ZACHARY J. KASTENBERG, AND MAX R. LANGHAM

INTRODUCTION

Treatment of the most common malignant liver tumor in children, hepatoblastoma (HB), has been one of the success stories of modern pediatric oncology. From a disease with survival rates of only 35% in the 1970s, overall survival now approaches 90% for patients with low- and standard-risk tumors, and 45–79% for patients with metastatic disease. This progress has been due to improved surgical techniques, including liver transplantation, and chemotherapy based upon refined risk stratification and treatment protocols. In addition, sharing of accumulated experience through improved international collaboration has led to a new global risk stratification system for HB using PRETEXT/POST-TEXT groups and annotation factors, age, and alpha-fetoprotein (AFP) level at diagnosis to assign risk-based treatment regimens. Pathologists in this international collaboration have established a new histopathological consensus classification for pediatric liver tumors. Concomitantly there have been advances in chemotherapy options, an increased rate of successful surgical resection for large bulky tumors, and progress in identifying biologic markers that portend increased or decreased risk.

Complete surgical resection remains a cornerstone of successful outcomes and modern liver resection requires a fundamental understanding of liver anatomy, sophisticated radiographic imaging, thorough knowledge of disease pathophysiology and treatment protocols, and refined surgical techniques, all performed within the support network of a multidisciplinary liver team.

DIFFERENTIAL DIAGNOSIS

Malignant liver tumors in children account for about 1.5% of all childhood malignancies. Hepatoblastoma (HB) is the most common, with a peak incidence between birth and 4 years of age. Hepatocellular carcinoma (HCC) is most often seen in older children and adolescents (**Table 58.1**). Other rare pediatric hepatic malignancies include malignant rhabdoid tumor of the liver in infants (<1 year), biliary rhabdomyosarcoma (3–4 years), and undifferentiated embryonal sarcoma of the liver (UESL) (6–10 years). Occasional tumors are seen that have features of both HB and HCC and used to be called transitional tumors. Their remarkable heterogeneity has begun to be recognized in recent years and they are now referred to as hepatocellular neoplasm-not otherwise specified (HcN-NOS), or "HB-with-HCC-features." By convention, these histologically hybrid tumors are treated as HBs.

Table 58.1 Age at presentation in malignant and benign liver tumors

Age group	Malignant	%	Benign	%
Infant/toddler	Hepatoblastoma (including hepatoblastoma with HCC features (HcN-NOS)[a]	40–50	Infantile hepatic hemangioma	10–20
	Rhabdoid tumor	1	Mesenchymal hamartoma	<10
	Others	1	Others	1
School age/adolescent	Hepatocellular carcinoma (including fibrolamellar HCC)	20	Focal nodular hyperplasia and other nodules	3–5
	Sarcomas and other	7	Hepatic adenoma and other	1

Source: Von Schweinitz D. Management of liver tumors in childhood. *Semin Pediatr Surg* 2006; 15: 17–24.

[a] Hepatocellular neoplasm-not otherwise specified (HcN-NOS) includes transitional type tumors with features of both HB and HCC.

Hepatoblastoma

HB accounts for about 80% of the malignant tumors in children and the incidence has increased over the past two decades. PRE-Treatment EXTent of disease (PRETEXT) is a radiographic staging system used in multicenter research trials to determine prognosis and treatment, including recommended timing of surgical resection for HB. POST-Treatment EXTent of disease (POST-TEXT) refers to the extent of tumor following neoadjuvant chemotherapy prior to surgical resection.

Hepatocellular carcinoma

Only 0.5–1% of all HCC manifest before 20 years of age and more than two-thirds of these are in children over 10 years of age. Chemotherapy is less effective for HCC so primary complete tumor resection would be considered at diagnosis if resection is at all possible. Often, however, these tumors present with metastatic disease and, if there is no response to chemotherapy, palliative measures of local control such as chemo- or radioembolization can be considered.

Sarcomas

UESL is the most common sarcoma of the liver in children and is thought to occur on a genetic continuum with its benign cousin, mesenchymal hamartoma. Most tumors present with cystic components and many have central areas of hemorrhage. If the tumor is not resectable at diagnosis, it is usually resectable after preoperative chemotherapy. Biliary rhabdomyosarcoma often presents with biliary obstruction and jaundice. The tumor is usually very sensitive to chemotherapy and radiation therapy. Angiosarcomas are exceedingly rare but have been reported as *de novo* tumors or following malignant transformation of an infantile hepatic hemangioma (IHH).

Rare malignancies and metastases

Malignant rhabdoid tumor of the liver may be confused with the small-cell undifferentiated variant of HB but INI1 tumor marker will be negative in rhabdoid tumor. Other rare but potential primary malignancies include germ cell tumors and malignant stromal epithelial tumor. Additionally, neuroblastoma, Wilms' tumor, pancreatic, and neuroendocrine tumors may be metastatic to the liver.

Benign tumors

The two most common benign tumors are hemangiomas:

- *Infantile hepatic hemangioma* expresses GLUT2 and responds to treatment with propranolol.
- *Rapidly involuting congenital hemangioma (RICH)* presents as solitary large masses that involute relatively rapidly and do not express GLUT2 or respond to propranolol.

Diffuse infantile hemangioma may occasionally be quite large and, when symptomatic, contemporary treatment includes propranolol and treatment of congestive heart failure caused by intratumoral arteriorvenous shunting; surgery and/or embolization is rarely necessary. Other benign tumors include mesenchymal hamartoma, biliary cystadenoma, hepatic adenoma, focal nodular hyperplasia (FNH), macroregenerative nodules, dysplastic nodules, and pseudoinflammatory myofibroblastic tumors.

Non-neoplastic masses such as vascular malformations, congenital and acquired cysts, abscess, hematoma, and fatty infiltration of the liver may occasionally be confused with liver tumors. Hepatic hematoma or infarction should be suspected in any child with a history of hepatic trauma or in newborns with sepsis and coagulopathy, especially if there is a history of perinatal birth trauma, thrombocytopenia, or hemodynamic collapse requiring cardiopulmonary resuscitation. Congenital liver cysts are rare and represent a spectrum ranging from large simple cysts to intrahepatic choledochal cysts and ciliated hepatic foregut cysts. Acquired cysts might be bacterial, hydatid, amoebic, or fungal abscesses.

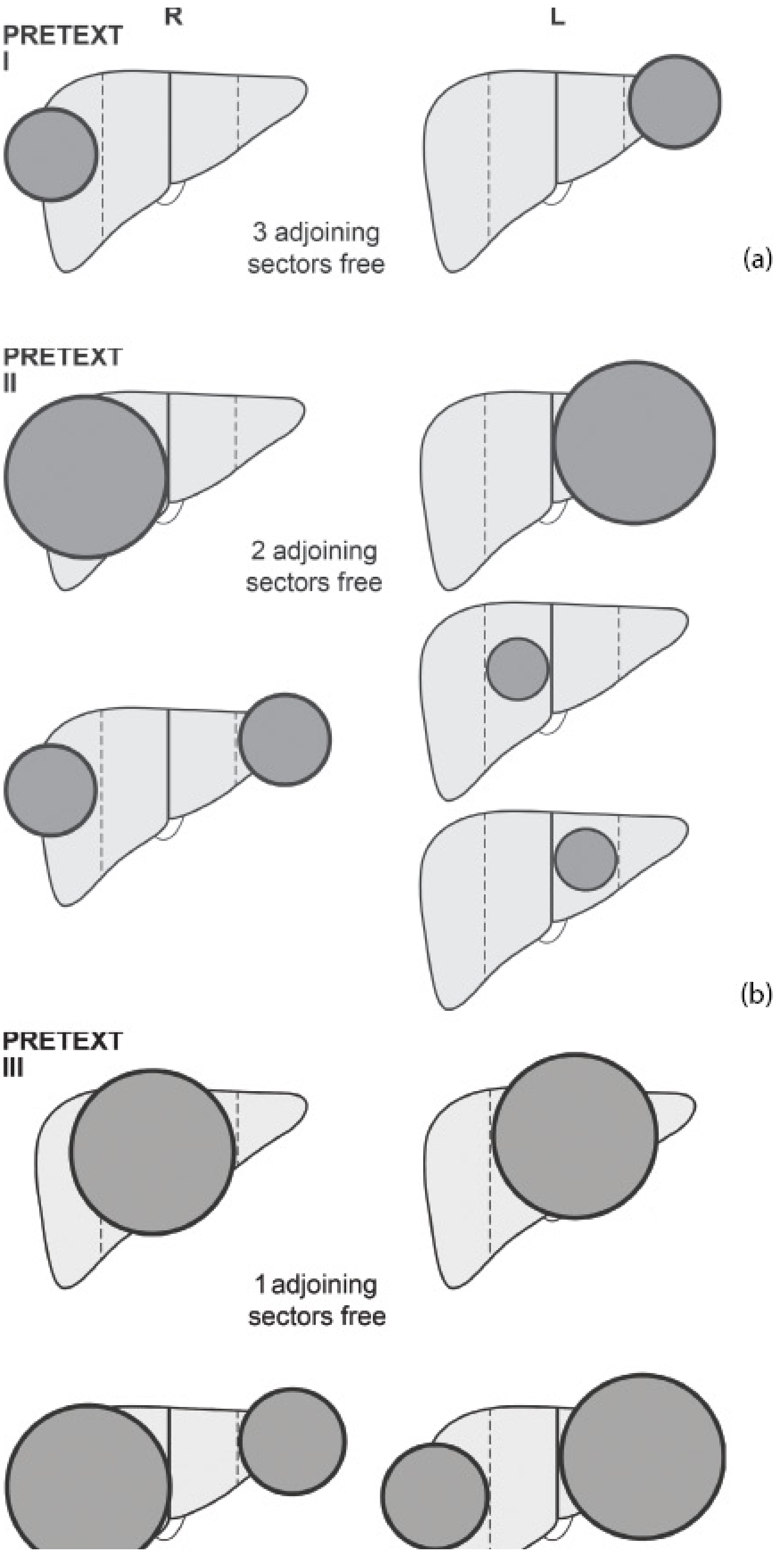
MULTIDISCIPLINARY PEDIATRIC LIVER TEAM

An experienced multidisciplinary team is the key to successful treatment of complex liver tumors. Such a team can reinforce and cross-check each other's performance to make evidence-based treatment choices and prevent mistakes. Team members will include an experienced pediatric anesthesiologist, oncologist, gastroenterologist, pathologist, radiologist, interventional radiologist, and surgeon with experience in complex liver surgery and, ideally, the availability of a surgeon with pediatric liver transplant experience. It is very important that the surgeon has a realistic understanding of their team as well as his or her own abilities in planning a major liver resection and/or transplant in a child. Referral to an adult hepatobiliary team is *not* a substitute for pediatric-specific knowledge and may result in surgical treatment decision-making that lacks insight into the major differences in biologic behavior, and surgical treatment algorithms between HB and HCC. Most liver resections performed in children are elective, but optimal outcomes depend on coordination of multidisciplinary therapy with relatively tight time windows. Avoidance of unnecessary surgical delays will facilitate adherence to evidence-based chemotherapy regimens thereby improving chances for long-term survival.

PREOPERATIVE CONSIDERATIONS

Presentation

Initial detection of a liver tumor is most often an abdominal mass palpated on well-child examination followed by a screening ultrasound showing a liver mass (**Figure 58.1**). AFP is almost always elevated in HB.


R
L
PRETEXT
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(b)
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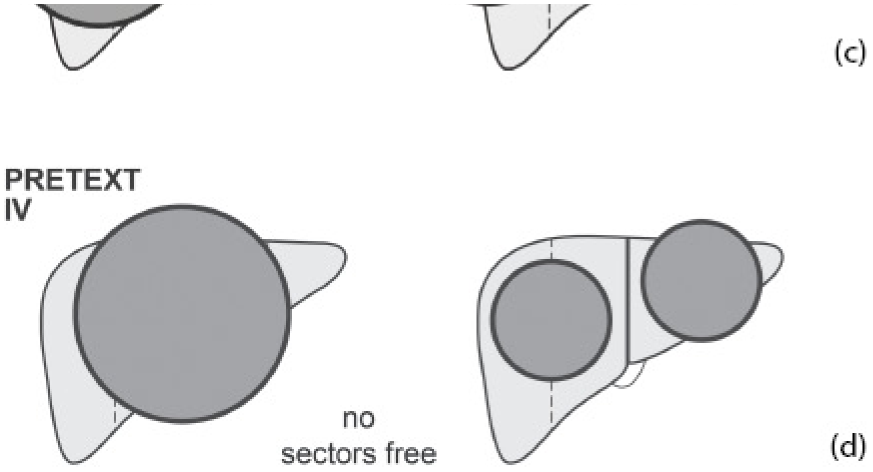
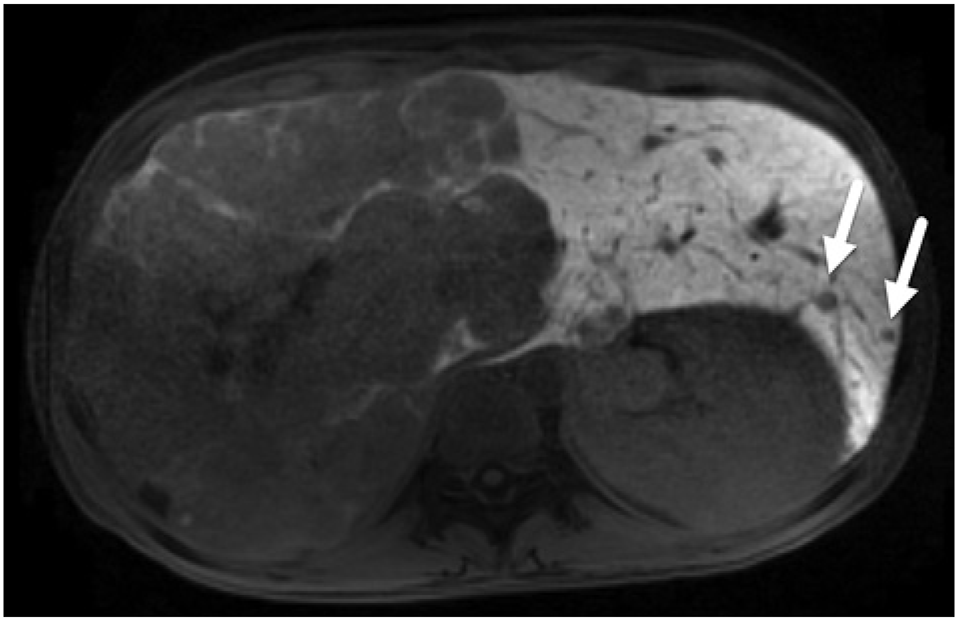


58.1

Imaging

Ultrasound (US) is the preferred initial imaging confirming the liver as the organ of tumor origin. Definitive radiographic imaging involves contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) of the abdomen and a chest CT to exclude metastatic disease in the lungs. The abdominal CT must include high-resolution images obtained in the venous phase to identify tumor proximity to the major portal and hepatic veins and inferior vena cava (IVC). Although some prefer the use of CT in the youngest patients because increased speed of CT image acquisition compared with MRI provides images with less motion artifact without sedation and anesthesia, others feel that MRI is the cross-sectional modality of choice even if sedation or anesthesia is required. They cite the lack of ionizing radiation and superior soft-tissue contrast resolution. MRI is ideally performed with hepatocyte-specific contrast agents that enhance the radiologist's ability to identify and diagnose small multifocal tumor foci that can be missed with either CT or MRI with routine gadolinium contrast (**Figure 58.2**). Currently, there are two hepatocyte-specific contrast agents on the market: Gadoxetate disodium (Gd-EOB-DTPA, Eovist/Primovist; Bayer, Leverkusen, Germany) and gadopentetate dimeglumine (Gd-

BOPTA, MultiHance; Bracco, Milan, Italy). Each agent has advantages and disadvantages.



58.2

PRETEXT/POST-TEXT and risk-stratified treatment in hepatoblastoma

In North America, HB disease stage was historically defined by a surgery based staging system:

- *Stage I:* Complete macroscopic resection at diagnosis
- *Stage II:* Macroscopic resection at diagnosis with microscopic residual disease
- *Stage III:* Macroscopic total resection with lymph node involvement or tumor spill/rupture or incomplete resection with macroscopic residual
- *Stage IV:* Metastatic disease

This system has been replaced by a PRETEXT based risk stratification that includes PRETEXT group, PRETEXT annotation factors VPEFR and M, patient age, and AFP level. This is both more accurate and applicable to the majority of HBs now treated with neoadjuvant chemotherapy. Image-defined risk factors are determined using the PRETEXT system, which is made of two components: the PRETEXT group and the PRETEXT annotation factors (see **Figure 58.1**; **Table 58.2**). The North American Children's Oncology Group (COG), along with its international colleagues from Europe and Japan, has adopted a new global risk stratification system for HB. Treatment assignments into very low, low, intermediate, and high risk treatment groups are based upon the PRETEXT group (I, II, III, IV), PRETEXT annotation factors (V, P, E, F, R, M), patient age (0–3, 3–8, or >8 years), and AFP level (<100, 100–1000, >1000 IU/L) at diagnosis. Approximately 80% of patients are best treated with upfront chemotherapy due to factors increasing the risk of a primary resection or the presence of metastatic disease, 60% low or intermediate risk, and 20% high risk. The final 20% of patients (very low risk) have PRETEXT group I or II tumors with negative annotation factors for which resection *at diagnosis* is recommended.

Table 58.2 **PRETEXT/POST-TEXT groups I, II, III, IV and annotation factors (V, P, E, F, R, C, N, M)**

Definition	
PRETEXT[a] / POST-TEXT group[b]	
I	One section involved Three adjoining sections are tumor-free
II	One or two sections involved Two adjoining sections are tumor-free
III	Two or three sections involved One adjoining section is tumor-free
IV	Four sections involved
PRETEXT/ POST-TEXT annotation factors	
V	Venous involvement, V, denotes vascular involvement of the retrohepatic vena cava or involvement of *all three* major hepatic veins (right, middle, and left)
P	Portal involvement, P, denotes vascular involvement of the main portal vein and/or BOTH right and left portal veins
E	Extrahepatic involvement of a contiguous structure such as the diaphragm, abdominal wall, stomach, colon, etc.
F	Multifocal tumor nodules
R	Tumor rupture at diagnosis
C	Caudate lobe
N	Lymph node involvement
M	Distant metastatic disease (usually lungs, occasionally bone or brain)

[a] PRE-Treatment EXTent of disease (PRETEXT).

[b] POST-Treatment EXTent of disease (POST-TEXT). Post-treatment is determined by radiographic imaging after chemotherapy but before surgical resection.

Multidisciplinary treatment of hepatoblastoma

Treatment of HB involves a combination of surgery and chemotherapy. The two most powerful chemotherapy agents used are cisplatin and doxorubicin. Other agents variably used by the different trial groups have included 5-flourouracil, vincristine, irinotecan, temsirolimus, pirarubicin, carboplatin, and etoposide. In general, chemotherapy is given until surgery can be performed on the primary tumor as well as any remaining detectable extrahepatic disease, and then continued postoperatively, typically with a total of six cycles of chemotherapy lasting 4–5 months. Common chemotherapy side effects include fever, neutropenia, infections, cisplatin ototoxicity, and, less commonly, kidney toxicity, cardiomyopathy, secondary malignancies, and infertility.

Cooperative multicenter trials based on risk-adapted medical and surgical therapy conducted by groups in Europe (International Childhood Liver Tumors Strategy Group/SIOPEL and German Society for Pediatric Oncology and Hematology [GPOH]), America (COG) and Japan (Japan Children's Cancer Group [JCCG]) have led to substantial advances in survival. Although different groups have historically defined their risk categories in disparate ways, survival of patients with low- and standard-risk tumors in all of these trial groups now approaches 90%.

Survival expectation of HB presenting with metastatic disease

Survival expectation for HB patients presenting with metastatic disease has improved significantly over the last 30 years (**Table 58.3**).

Table 58.3 Historic survival expectation of HB presenting with metastatic disease

Trial	Years	Percentage survival (%)
SIOPEL I	1990–1994	28
SIOPEL II	1994–1998	44
SIOPEL III-HR	1998–2006	56
SIOPEL IV	2005–2009	79

Experience within COG supports the efficacy of definitive resection at diagnosis for low-risk tumors with no or reduced chemotherapy following resection. Chemotherapy used either after resection of small standard histology tumors or before resection of larger, more complicated tumors is cisplatin/5FU/vincristine/doxorubicin (C5VD). This combination has good efficacy and a reasonable toxicity profile. For unresectable tumors in patients with intermediate-risk disease, liver transplantation has resulted in excellent survival and quality of life. There is much less sophisticated information on combination chemotherapy for the malignant tumors other than HB and protocols for these tumors have not substantially improved survival (see "Further reading").

Transatlantic differences

Complete local control of primary and metastatic disease remains the most critical intervention required to achieve long-term cure of malignant liver tumors. An historic defining difference between the American/COG and European/SIOPEL approaches has been an attempt at surgical resection at diagnosis when feasible in COG versus preoperative chemotherapy for all in SIOPEL. In the new Pediatric Hepatic International Tumor Trial (PHITT), more recent collaborative international surgical resection guidelines are now the same in Japan, North America, Australia, and Europe and rely upon PRETEXT and POST-TEXT to assist the individual surgeon in determining if upfront resection should be considered. In these guidelines, resection at diagnosis is recommended when a sectorectomy or simple (not extended) hemihepatectomy will predictably yield a complete R0 resection (i.e. localized PRETEXT group I or II tumors with negative PRETEXT annotation factors). A minimum 1 cm margin is recommended for *resection at diagnosis*; however, the 1 cm margin may not be as important when resection is performed *after chemotherapy* if the chemotherapy response has been robust. For extensive multifocal tumors, or any tumor with persistent bilobar major vascular involvement after chemotherapy (positive V or P on POST-TEXT imaging), referral to a specialty liver center for either radical resection or liver transplant is recommended. The improved surgical resection rates now possible for large complex tumors have contributed significantly to the improved outcomes achieved in recent trials. Lung metastases should be resected when they fail to clear with chemotherapy, although the role of metastasectomy is less clear for relapse or progressive disease in the lungs. These patients should be treated on a case-by-case basis.

BIOPSY AT DIAGNOSIS FOR UNRESECTABLE TUMOR

Tumor biopsy at diagnosis will depend upon the suspected tumor type. For HB, resection at diagnosis is recommended for PRETEXT I and many

PRETEXT II tumors. Biopsy, not resection, at diagnosis is recommended under the following circumstances:

- PRETEXT II with <1 cm radiographic margin on the middle hepatic vein and/or portal bifurcation
- PRETEXT III
- PRETEXT IV
- Any tumor with positive annotation factors (V+, P+, E+, F+, R+, C+, N+, and/or M+)

The type of biopsy procedure performed — open versus laparoscopic versus percutaneous — will be at the discretion of the treating institution. In general, however, many centers increasingly prefer using an image-guided, coaxial core needle biopsy (CNB) with embolization of the tract at procedure completion. CNB is particularly advantageous in the case of HCC where minimizing spill and contamination by thorough tract embolization is important.

- Ideal core needle approach would be through a buffer zone of overlying adjacent normal liver that will be eventually be included in the definitive surgical resection. Biopsy approach should not result in contamination of liver that will not ultimately be resected as this could result in a remnant of liver which contains a tumor-seeded biopsy tract.
- The recommended amount of tumor tissue to be retrieved is 7–13 cores, comprising 2 or 3 cores of viable tumor for diagnostic purposes, and 5–10 cores of viable tumor to be frozen for biologic studies. CNB is done under US guidance, avoiding adjacent vascular structures and sampling different areas of the tumor. Proper tissue assessment for viable tumor and handling require immediate tissue analysis by the pathologist.
- The recommended amount of normal liver to be retrieved for diagnostic purposes and for biologic studies is 1 or 2 cores. This is to rule out underlying liver disease and assess germline mutation, and these cores should be annotated and frozen with the biology samples for future biologic analysis.

TIMING AND TYPE OF SURGICAL RESECTION

The multidisciplinary treatment protocol will usually recommend the timing and type of surgical resection based upon the protocols surgical resection guidelines. These guidelines are heavily dependent upon the PRETEXT and POST-TEXT assessment of the imaging.

Hepatoblastoma

HB is often exquisitely chemosensitive. For initially unresectable tumors, with the possible exception of an extensive multifocal tumor, a decision about tumor resectability versus transplant will be made after assessing the initial response to chemotherapy. Continuing chemotherapy in a vain attempt to render an unresectable tumor resectable must be strictly avoided. Prolonged courses of preoperative chemotherapy (more than four cycles) can induce drug-resistant gene expression thereby selecting out surviving drug-resistant tumor clones. This has been hypothesized to increase the risk of failure and relapse. Most of the response to chemotherapy will occur with the first few cycles, with a diminishing response for each additional cycle. When not resectable at diagnosis, current protocols may recommend surgical resection or transplant, after either two, three, or four cycles. Plan the anticipated date of surgical resection far in advance, review the PRETEXT and POST-TEXT images with the liver multidisciplinary team, and reserve an appropriate block of operative time. Consult a liver specialty surgical team and/or transplant team for patients where the residual liver volume would be too small, and/or the major vascular involvement too extensive, for patient safety and where a transplant allograft would prevent leaving residual disease. Early referral to allow listing and waiting time is very important.

Hepatocellular carcinoma and rhabdoid tumors

These are less chemosensitive than HB and in general complete R0 resection is necessary for cure. However, some HCC in children may have heterogeneous histology, HB with HCC features, and these children are treated according to HB (not HCC) protocols.

Sarcomas

This group includes undifferentiated embryonal sarcoma of the liver (UESL), biliary rhabdomyosarcoma (RMS), and angiosarcoma. These have varying levels of chemosensitivity and radiosensitivity and treatment protocols must often be individualized based upon initial PRETEXT and POST-TEXT imaging and response to neoadjuvant therapy. The results of UESL and biliary RMS are approaching the survival rates achieved in HB.

Benign tumors

Some benign tumors can be safely observed if asymptomatic, and if malignancy can be definitively ruled out (i.e. IHH, some FNH, etc.). With advances in hepatocyte-specific contrast MRI imaging, malignancy may be ruled out radiographically, although close follow-up is needed, and errors in diagnosis continue to occur. When there is any question, biopsy or resection may be needed to definitively rule out malignancy. Mesenchymal hamartoma occurs on a genetic spectrum with UESL and surgical resection is generally recommended. Resection versus observation of hepatic adenomas may often depend upon genetic analysis of a percutaneous biopsy, or the presence of a known germline mutation.

PREOPERATIVE CHECKLIST

Preoperative evaluation should include the following factors:

- *Neutropenia/thrombocytopenia.* Post-chemotherapy marrow suppression can be assessed several days before surgery; absolute neutrophil count (ANC) of <1 may benefit from granulocyte colony stimulating factor (GCSF) given several days before surgery.

- *Preoperative laboratory profile* should include white blood cell (WBC) count, hematocrit, platelet count, ANC, electrolytes, lactate, prothrombin time/INR (PT/INR), aspartate transaminase/alanine transaminase (AST/ALT), fractionated bilirubin.
- *Blood products.* Type and cross 20 mL/kg packed red blood cells, 20 mL/kg frozen plasma, 10 mL/kg platelets. Many hospitals will have protocols for oncology patient-compatible blood products.
- *Consent, education, and emotional condition of patient and family.* It is important to consider the education and emotional state of the family and child. Reviewing in some detail the imaging studies with them, and clearly describing the steps of the operation, is helpful to the surgeon as well as the family. After the technical aspects of the operation have been covered and consents signed, make sure you take time to discuss emotional fears and goals with the parents. Letting them know there will be a comfortable area in which to wait, a clear communication plan for getting them updates on their child's condition and any changes in the operative plans is important. A few minutes spent focused on their comfort and well-being will be deeply appreciated and long remembered.

ANESTHESIA ISSUES AND CONCERNS

- *Intravenous access and monitoring.* Central venous catheter for monitoring and for fluid resuscitation and two large bore peripheral catheters. Groin or lower extremity lines should be avoided due to potential need for intraoperative clamping of the IVC. An upper extremity arterial line is highly recommended for continuous arterial pressure monitoring.
- *Pre-incision antibiotics,* first-generation cephalosporin,should be administered <60 minutes before incision.
- *Blood products.* Cross-matched, oncology patient-compatible, packed red blood cells (20 mL/kg), fresh frozen plasma (20 mL/kg), and platelets (10 mL/kg) available. Blood loss in a typical hepatic resection is much higher than in other operations performed by pediatric surgeons and the entire

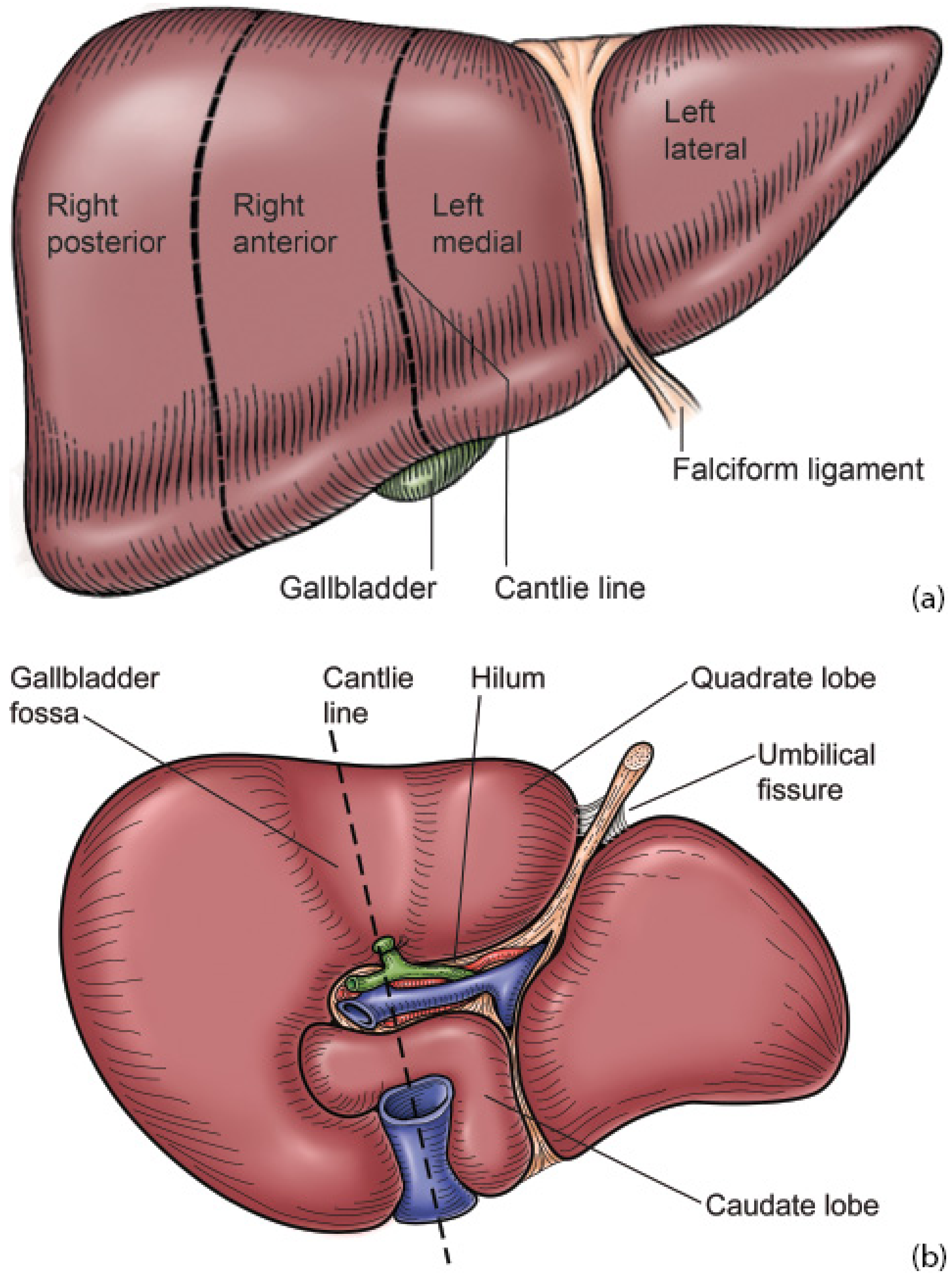
surgical team must be aware of this fact and feel comfortable participating in the operation.

- *Hypovolemic resuscitation.* The principle of hypovolemic anesthesia during the parenchymal division phase of complex liver resection, followed by intraoperative volume resuscitation is of real value and surgeons should discuss this strategy with the anesthesiologist preoperatively. Limiting volume resuscitation during the parenchymal transection phase of the operation by maintaining a low central venous pressure is important, if not critical, to the success of the operation and maintenance of patient safety. Hypotension is a routine part of the operation, particularly during partial compression of the vena cava, and may require administration of vasopressor support. Aggressive volume-loading causes hepatic congestion that increases the chance of bleeding from multiple veins simultaneously and can lead to a disastrous destabilization. Urine output during the hypovolemic resection is often quite low and will resume with post-resection volume resuscitation.
- *Air embolization.* During uncontrolled bleeding from the IVC or large hepatic veins air embolization is possible and is a potential cause, along with concomitant hypovolemia, of intraoperative cardiac arrest. End tidal CO_2 will be lost and the anesthesiologist must be aware of and have a treatment plan in place for this possibility. Rapid placement in Trendelenberg position, with the table rolled so the patient's right side is up, along with effective CPR often is rewarded by rapid return of stability as long as source control for the air and blood loss is achieved. While CO_2 embolization is usually better tolerated, arrests have been documented during laparoscopic liver resection. In this situation, reducing the intra-abdominal pressure is an important part of the resuscitation.
- *Hypothermia* can be more of a problem in children than in adults. Active measures for warming children include warming blood products, warmed forced air under the patient, and keeping the operating room temperature higher than may be comfortable for the surgical team.

SEGMENTAL ANATOMY

Segmental anatomy defines the basis for anatomic liver resection. Ideally, a liver resection will include resection of the complete segment or sector in continuity with its unique inflow and outflow.

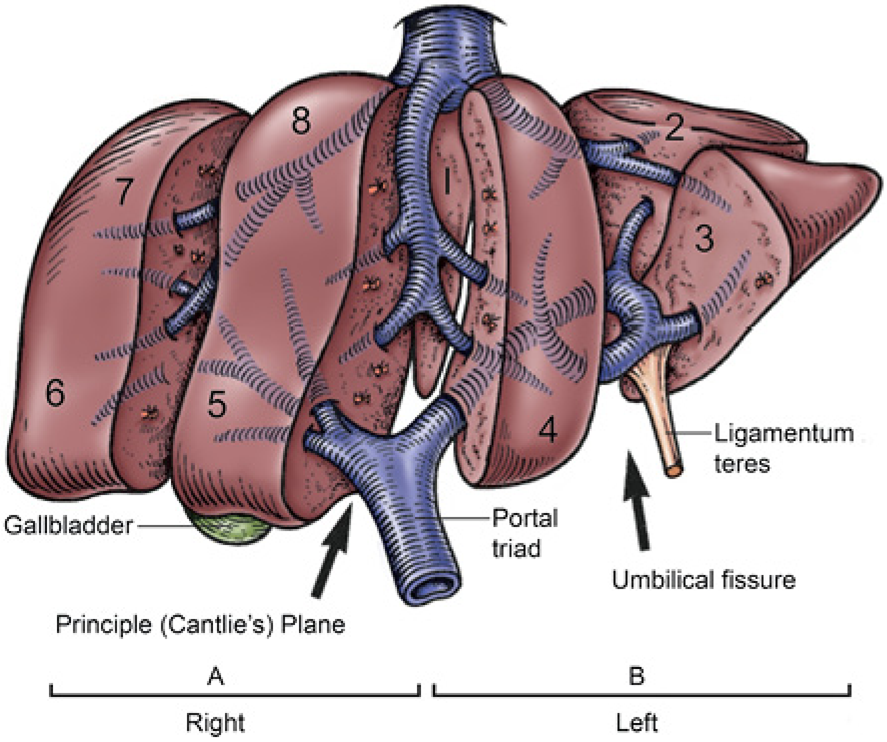
The right and left lobes (or hemilivers) each have two sectors. The ligamentum teres is the remnant of the fetal umbilical vein. It runs from the umbilicus within the inferior edge of the falciform ligament, through the umbilical fissure, where it becomes the left portal vein in the "Rex" recessus. The falciform ligament extends to the diaphragm, where it is continuous with the long left triangular ligament fixing the left lobe to the left diaphragm. On the right side it is contiguous with the coronary ligaments and short right triangular ligament defining the much broader "bare area" of the liver. Demarcation between left and right lobes is actually an imaginary line (Cantlie's line) which runs from the gallbladder fossa back to the retrohepatic vena cava; the middle hepatic vein lies in this plane. The left lobe (40% by volume) is divided into a lateral sector and a medial sector by the umbilical fissure. The right lobe (60% by volume) is divided into an anterior sector and posterior sector by the right hepatic vein (**Figure 58.3a and b**).


Right posterior
Right anterior
Left medial
Left lateral
Falciform ligament
Gallbladder
Cantlie line
(a)
Gallbladder fossa
Cantlie line
Hilum
Quadrate lobe
Umbilical fissure
Caudate lobe
(b)

58.3a,b

The liver is further divided into eight anatomic segments (Couinaud classification) (**Figure 58.4**). Each segment is supplied by a portal pedicle and each drains into a tributary of a hepatic vein. Segment 1 is the caudate lobe and is supplied by branches from the posterior aspect of right and left portal veins with venous drainage directly into the retrohepatic IVC.

Segments 2 (superior aspect) and 3 (inferior aspect) comprise the left lateral sector; segment 4 is the left medial sector. Segment 4 can also be subdivided into 4a (superior) and 4b (inferior). Segments 5 and 8 form the right anterior sector; segments 6 and 7 form the right posterior sector.


8
7
1
2
3
6
5
4
Ligamentum teres
Gallbladder
Portal triad
Umbilical fissure
Principle (Cantlie's) Plane
A
Right
B
Left

58.4

TYPES OF LIVER RESECTION

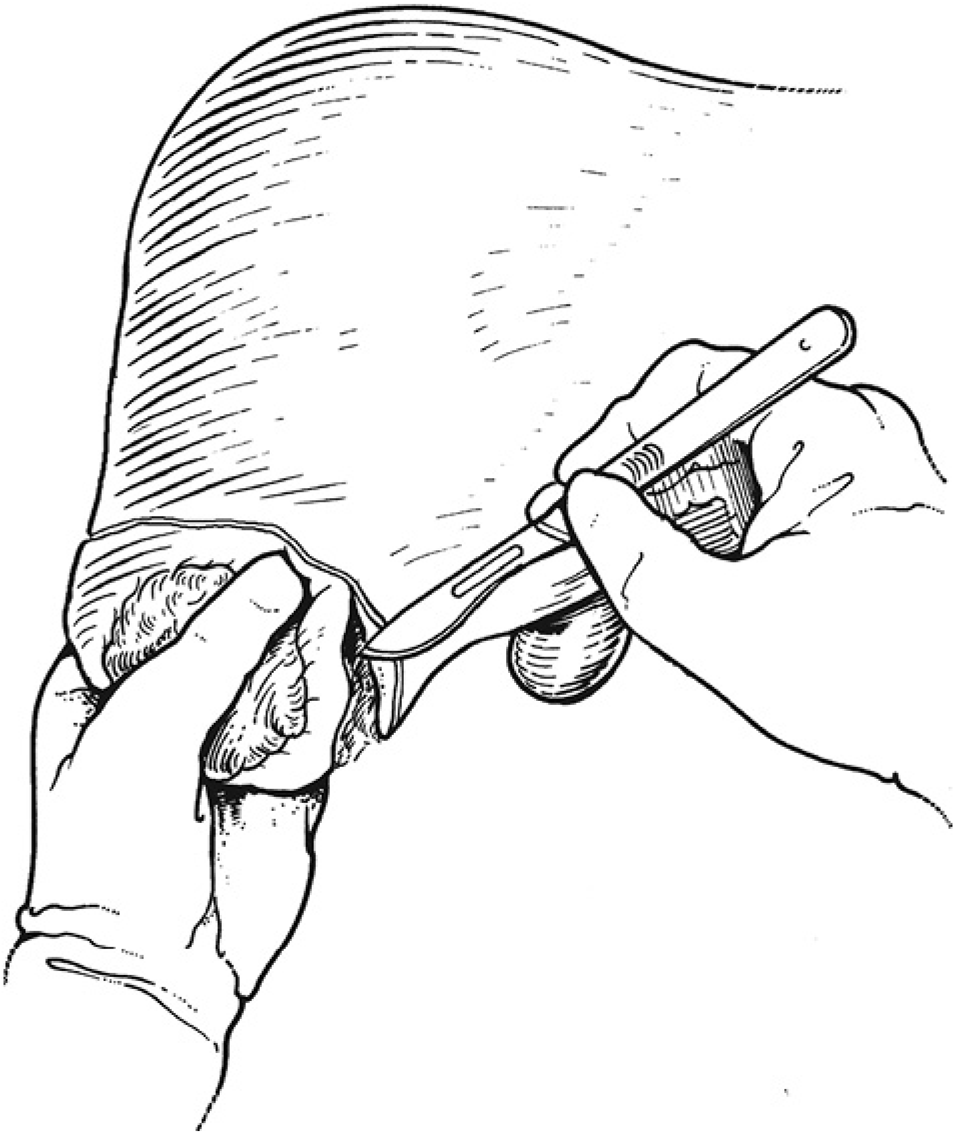
The nomenclature of liver resections has changed over the years. Listed here is the current consensus classification used by most specialty liver surgeons (see Strasburg et al. in "Further reading").

- *Left hemihepatectomy* involves resecting segments 4a, 4b, 2, and 3 with or without portions of segment 1. Preservation of the entire right portal vein, the arterial supply to segments 5, 6, 7, 8, and the bile ducts draining the right liver and the right hepatic vein is essential.
- *Extended left hemihepatectomy* involves resecting segments 4a, 4b, 2, and 3 with or without portions of segment 1, *plus* resection of all or part of

segments 5 and 8 with extension of the resection to the right of the middle hepatic vein.

- *Right hemihepatectomy* involves resecting segments 5, 6, 7, 8. Preservation of the entire left portal vein, the arterial supply to segments 2, 3, 4, and the bile ducts draining the left liver and the caudate and the left hepatic vein is essential.
- *Extended right hemihepatectomy* involves resecting segments 5, 6, 7, 8 *plus* resection of all or part of segments 4a and 4b with extension of the resection margin to the left of the middle hepatic vein.
- *Mesohepatectomy* is a central liver resection of segments 4a, 4b, 5, 8. It will involve dissection of bilateral portal, arterial, and biliary structures and careful attention must be given to preservation of inflow and outflow to the remaining left lateral and right posterior sectors. The choice of this resection in preference to a transplant or extended hemihepatectomy must be made by an experienced specialty liver team.
- *Sectoral resection or sectorectomy (alternatively sectionectomy).* Left lateral sector resection involves resection of segments 2 and 3. Right posterior sector resection involves resection of segments 6 and 7 with or without portions of segment 1.
- *Segmentectomy.* For resection of individual Couinaud segments, the isolation of the portal triads usually involves some intraparenchymal dissection. The inferior segment 3 on the left and the combined inferior segments 5 and 6 on the right are more amenable to an anatomic segment dissection. Single segment dissection for the superior segments 2, 4a, 7, and 8 are challenging and these procedures may resemble wedge resection. Isolated caudate (segment 1) resections are challenging since the caudate often gets portal supply from both the right and left branches of the portal vein and the biliary anatomy is highly variable. Multiple hepatic veins drain the caudate and a meticulous dissection of the retrohepatic vena cava is required.
- *Wedge resection* is a non-anatomic “wedge” resection around the tumor with a rim of normal hepatic parenchyma (**Figure 58.5**). Great care must be taken not to injure the blood supply and/or biliary drainage of the

remaining segments of liver. The advisability of a wedge resection, as opposed to a formal anatomic segmental resection, when performing surgery for cancer is debated and many feel that this should be done only in the setting of benign lesions where the risk of relapse is less important.



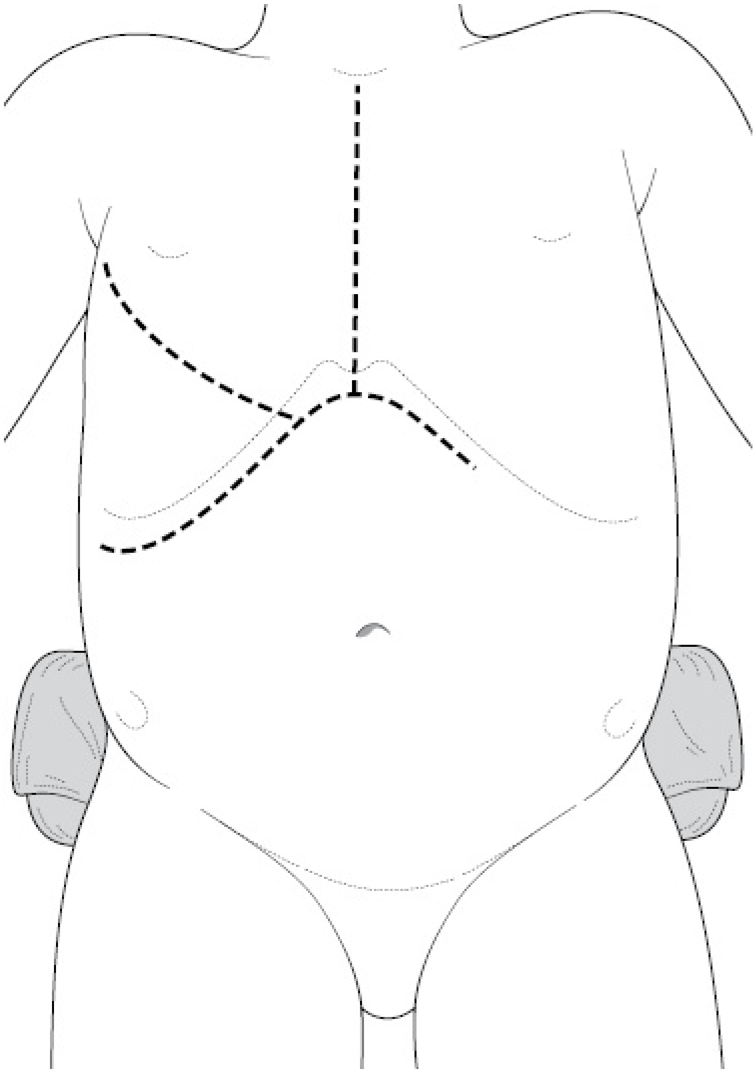
58.5

OPERATION

Safe resection requires detailed anatomic knowledge, comfort obtaining vascular control, and facility in multiple techniques of vascular reconstruction. The surgeon must be well-organized and calm and have the ability to expeditiously deal with bleeding while maintaining situational awareness about the patient's condition and anatomy, so as not to compromise the liver remnant or violate the tumor margin. A well-designed and executed hepatic resection removes the tumor with little or no physiologic impact on the child and a hospitalization that should be about a week in duration.

Incision

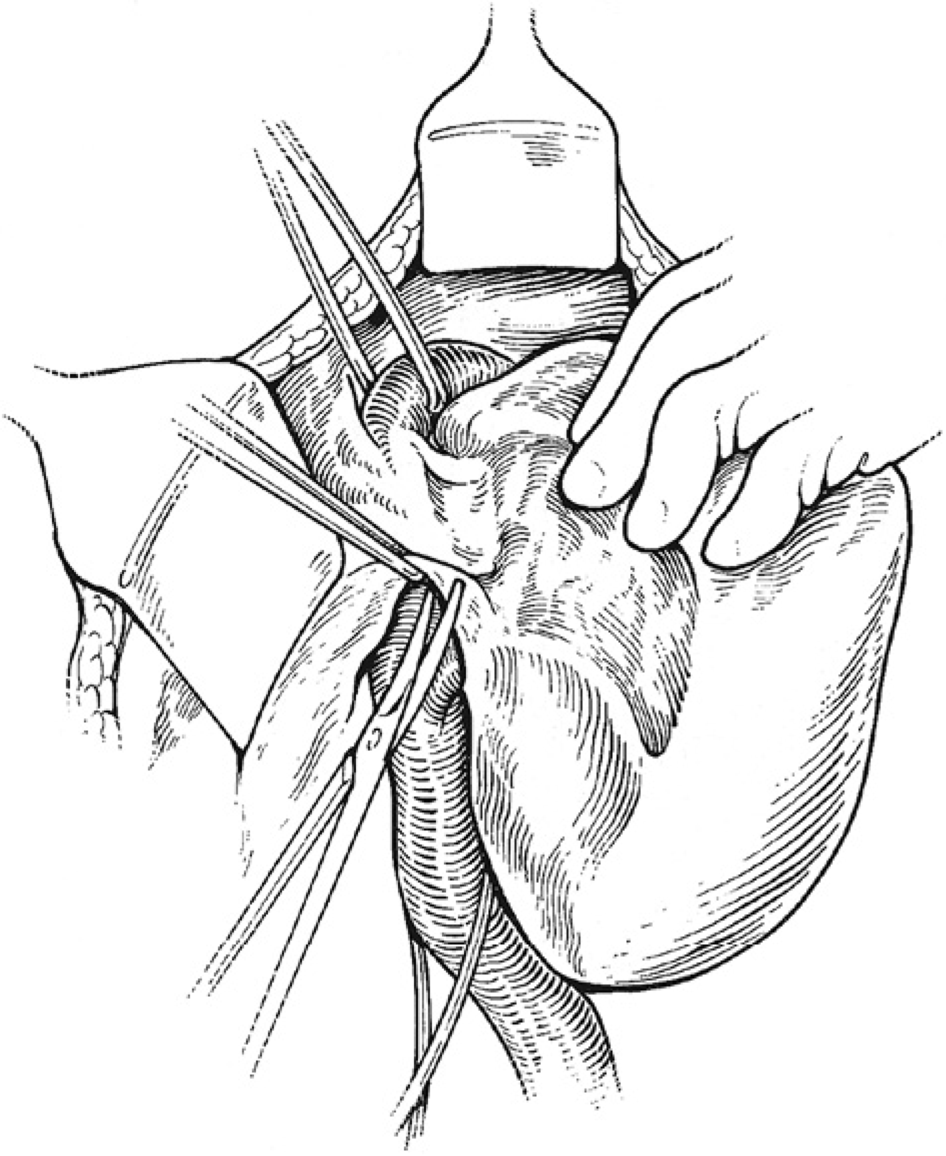
Large tumors require excellent visualization, and a Mercedes, or inverted Y, incision similar to the one used for liver transplantation is commonly used (**Figure 58.6**). Excision of smaller tumors on the right can often be performed through an incision beginning at the xiphoid and extending down the linea alba to a point 2–3 cm below the costal margin, then extending subcostal to the eleventh rib laterally. Left-sided resections can be performed through a subcostal chevron with or without a vertical extension to the xiphoid or a midline incision. If more extensive exposure of the suprahepatic vena cava is needed, a right thoracoabdominal incision can be helpful. Tumor rupture, either spontaneous or at the site of a previous biopsy, is not uncommon and may result in tumor adherence to the peritoneal surface of the abdominal wall and/or diaphragm. The surgeon should enter the abdominal cavity at a location remote from any anticipated area of tumor adherence, and the attached structures (abdominal wall, diaphragm, omentum, etc.) should be left attached to the tumor and excised en bloc.



58.6

Mobilization of liver

The initial maneuvers to mobilize the liver depend to some extent on the position of the tumor mass. Divide ligamentum teres and dissect the falciform ligament superiorly to the anterior surface of the suprahepatic vena cava usually for all tumors. Exposure is greatly facilitated by use of an open architecture retraction system such as the Thompson®, Omni®, or Bookwalter® retractor systems. The left lobe is freed from the diaphragm by dividing the left triangular ligament with exposure of the caudate lobe and left hepatic vein. The right lobe is mobilized off the diaphragm by dividing the superior and inferior coronary ligaments with exposure of the “bare area” and then retrohepatic cava (**Figure 58.7**). With smaller tumors, leaving some of the attachments in place on the uninvolved side may help to ensure that the remaining hepatic remnant will not be subject to torsion on its vascular pedicles.



58.7

Intraoperative decision-making

Few types of surgery are less forgiving of poor decision-making than the surgical resection of a large liver tumor in a small child. For this reason, it is important that the surgeon makes a habit of very deliberately taking stock of the information presented to them before proceeding to that portion of the operation from which there is no easy return. This point of no return is generally the ligation of either the inflow or outflow of a segment or sector of liver but ideally precedes any dissection which would make subsequent operations such as a liver transplantation more difficult. The following checklist is designed to help organize the data which inform this decision.

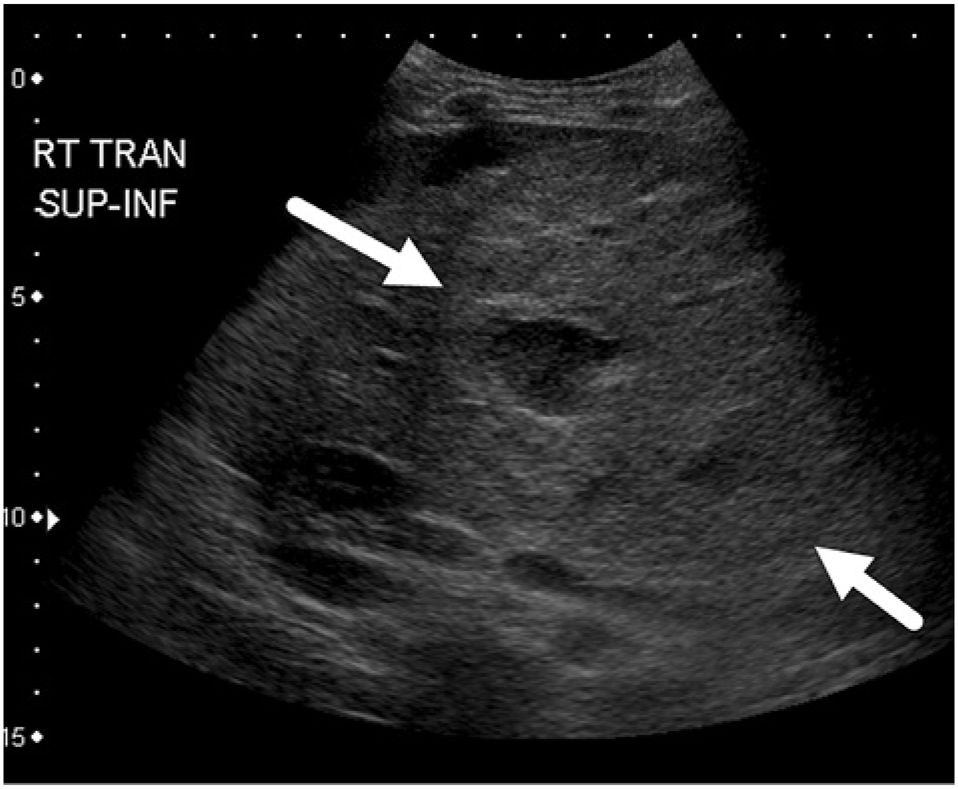
- What is the status of the liver parenchyma? Fibrosis, cirrhosis, and fatty or other underlying liver disease will increase the risk of postoperative hepatic insufficiency and may represent a contraindication to an anticipated small for size liver remnant.
- Is there extensive multifocal disease? This may be a contraindication for conventional resection unless liver transplant options have been discussed in detail and the team has specifically rejected the option of transplantation based upon uncontrolled extrahepatic tumor.
- Does the proposed liver remnant have an unanticipated focus of tumor, or is it otherwise compromised? Will the liver remnant be too small for size?
- Is an adequate margin of resection in doubt?
- What is the status of the vascular inflow and outflow to the proposed liver remnant? Assessment of the vascular anatomy should include an estimation of ischemic time, if any, needed for vascular reconstruction. Can this be done safely?
- Intravascular tumor thrombus. Trouble arises when there is tumor thrombus extending into the remnant liver hepatic vein or vena cava outflow, or portal venous inflow. This finding, ideally picked up on preoperative imaging, should prompt transplant consideration. Tumor thrombectomy of viable tumor may increase the risk of relapse. Resection of the portion of vein with adherent tumor thrombus and reconstruction

with the jugular vein or other conduit is possible but is an advanced technique that should not be undertaken casually.

- Is the entire operative team prepared for the resection required by operative findings? Does the institution have the resources to care for potential operative complications of the resection required?

Intraoperative ultrasound

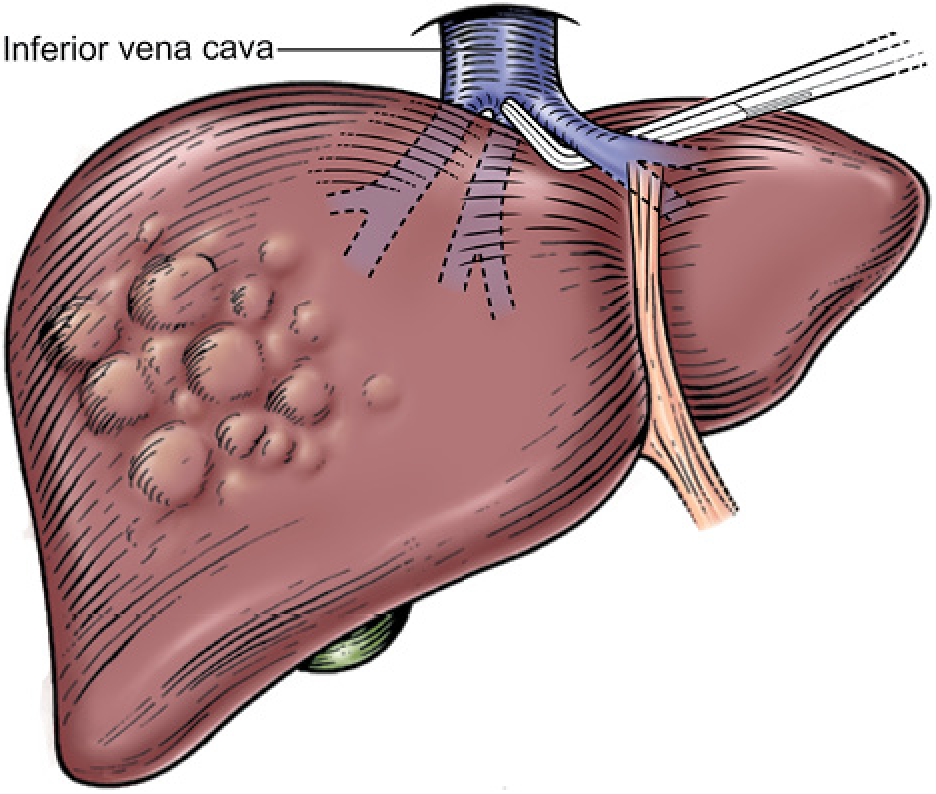
Intraoperative US of the liver before beginning vascular isolation or parenchymal dissection may be advisable in the setting of vascular involvement by the tumor or close margin (**Figure 58.8**). In a soft liver it may be easy to feel the solid tumor and judge parenchymal margins, but it is difficult to use tactile impressions to map the hepatic venous or portal structures and their relationship to the tumor edges. Preoperative imaging sometimes miscalculates the extent of disease because mass effect and compression by a large tumor can distort the anatomy. The surgeon, holding the liver and US probe in hand, has an enhanced ability to discern the true relationships of the tumor to the vessels, to plan resection margins, and to exclude multifocal disease in the planned remnant liver. Visualizing a three-dimensional image of the relationship of the tumor to the vessels takes some experience but is not particularly complicated if one has a solid knowledge of the segmental anatomy of the liver. The Glissonian capsule is quite bright (echodense) and portal structures can be distinguished from hepatic veins by the presence of the capsule around the former. Biliary obstruction and cirrhosis are distinctly uncommon in HB and cirrhosis is a contraindication to more than a trivial resection. Both of these entities dramatically change the US picture of the liver and are relevant in older patients with other problems such as HCC.



58.8

Suprahepatic vena cava and hepatic veins

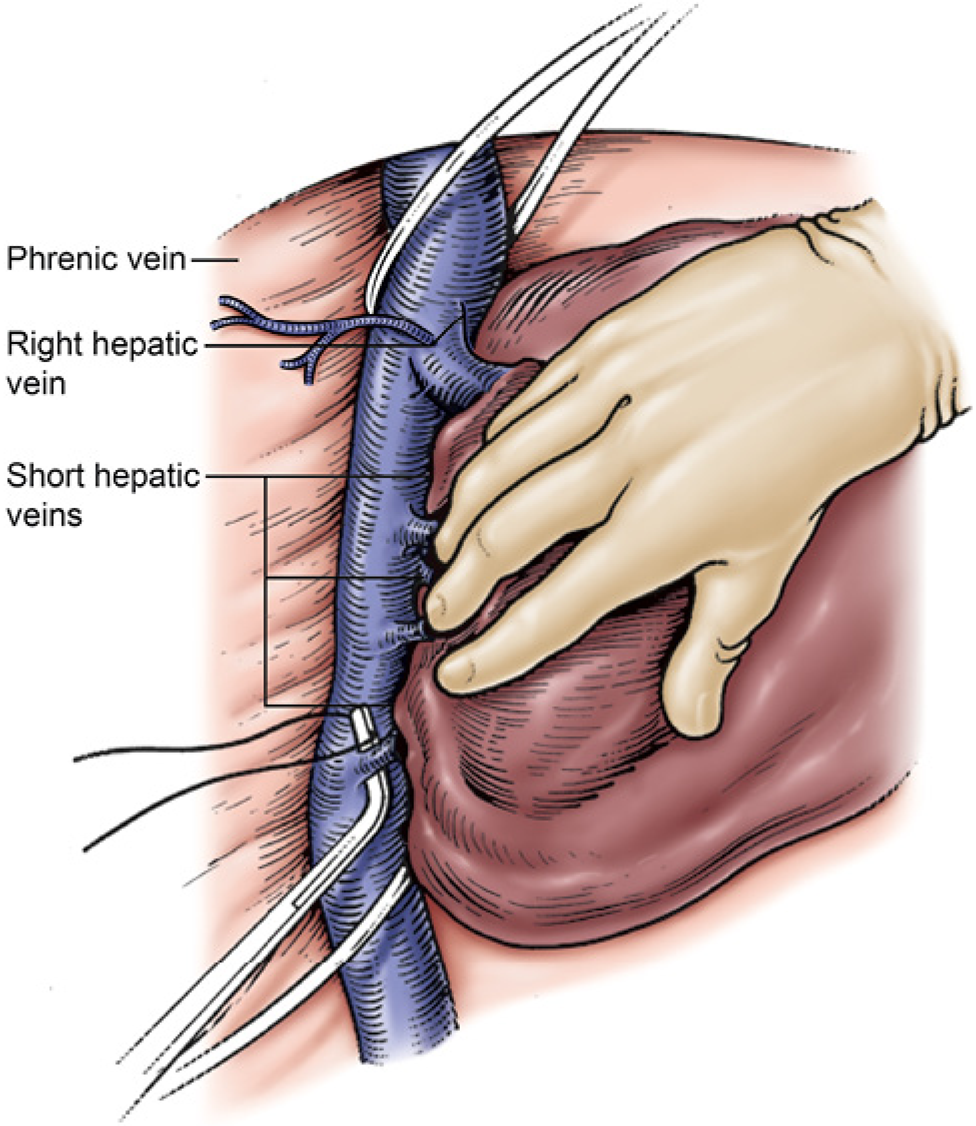
With mobilization of the left lateral sector, the left hepatic vein and caudate are exposed and dissected. Thick investing fascia of the diaphragm is carefully teased away to gently expose and mobilize the suprahepatic vena cava, left hepatic vein, and middle hepatic vein, which often share a common trunk (**Figure 58.9**). As the dissection is carried down the right lateral aspect of the suprahepatic vena cava, the right hepatic vein is cleaned and identified. On both sides phrenic veins are at risk and should be identified and protected or suture ligated. After dissecting both medially and laterally, the vena cava and dome of the liver can be picked up by the surgeon to facilitate the dorsal part of the dissection. This will partially occlude the venous return and allow the anesthesia team to judge the degree of hypotension to expect if the cava is clamped later in the case. The cava is freed from its dorsal attachments by gentle blunt dissection and either a vessel loop, or umbilical tape can be placed around the suprahepatic vena cava. Alternatively, once thoroughly mobilized and cleared, the suprahepatic vena cava control can be achieved with an immediately accessable Klintmalm™ or similar large gently curving vascular clamp. Tapes or vessel loops placed around the cava and/or hepatic veins should be loose at this point in the operation to prevent inadvertent obstruction, which can cause both unnecessary hypotension and venous engorgement of the liver and intestines. This limits exposure and can in extreme cases cause acidosis and bowel or liver injury.



58.9

Dissection of the retrohepatic vena cava

Resection of tumors involving Couinaud segments 6, 7, and sometimes 8 will benefit from a meticulous dissection of the retrohepatic vena cava to free the short hepatic vein attachments (**Figure 58.10**). If ties are used, care must be exercised in tying them or the tie should be reinforced with a Ligaclip®, as an increase in the central venous pressure can push off an imperfect knot and lead to significant blood loss. This is particularly true of the main right hepatic vein, which optimally is oversewn or stapled.



58.10

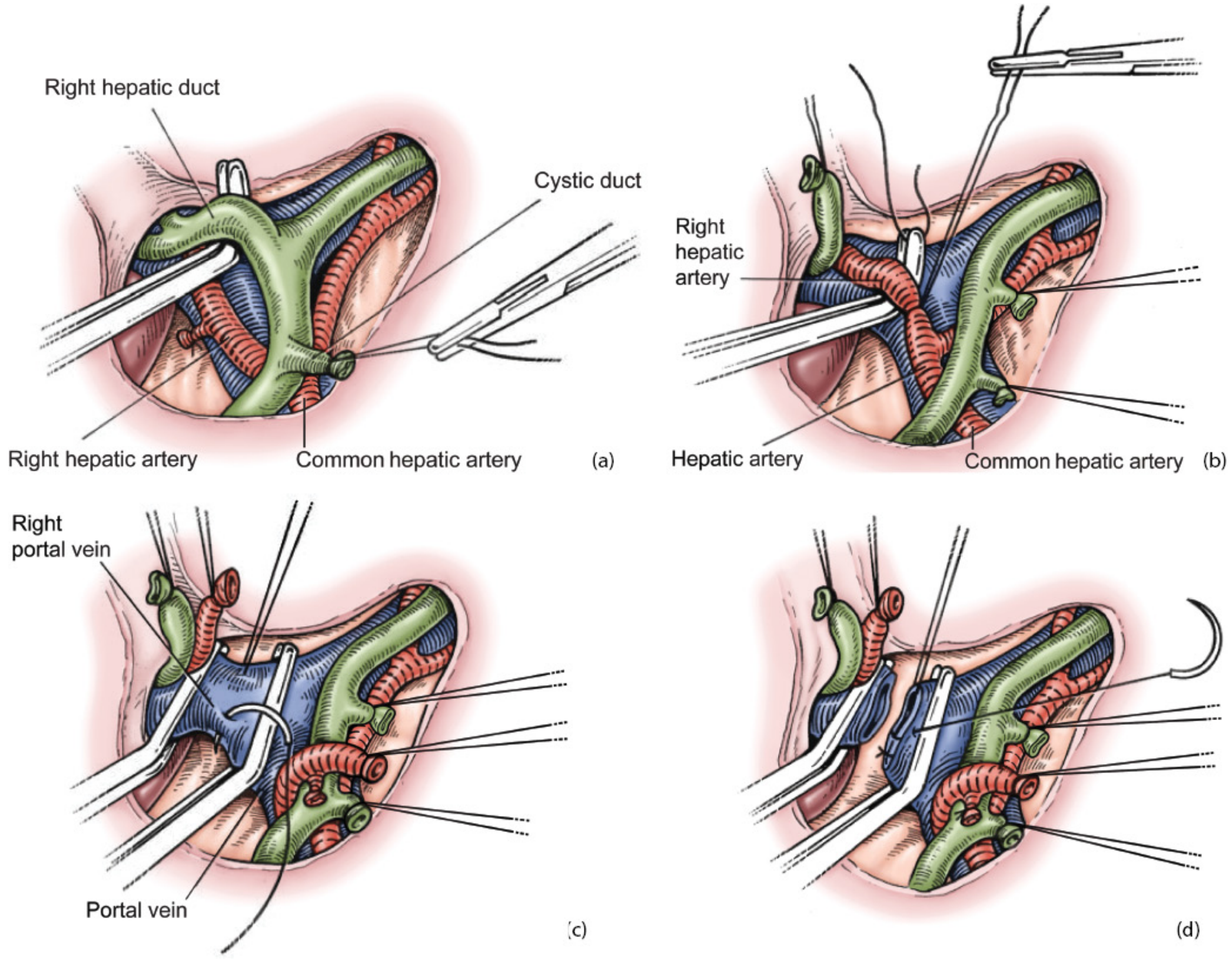
If the caudate lobe is to be mobilized off from the retrohepatic cava, the surgeon should be aware of, and prepare for, a particularly large caudate perforating vein, which is usually located about 1 cm inferior to the suprahepatic orifice of the left hepatic vein. This area should not be dissected until full mobilization has been achieved because inadvertent tear and bleeding from the caudate perforating vein can be challenging to control without good exposure.

Proximal and distal control of the supra- and infrahepatic vena cava should be planned during this phase of the dissection. This can be achieved either by preplacement of umbilical tape or vessel loops, or test clamping with confirmed exposure and preselected vascular clamps that are readily available on the field.

Drainage of Couinaud segment 6 is influenced by the presence, or absence, of an auxiliary inferior right hepatic vein. Knowledge of the presence or absence of this accessory vein is important during complex resections. If the vein is present, the surgeon may be able to preserve venous outflow of segments 5 and 6 and save these segments when the main right hepatic vein is compromised.

Exposure and control of porta hepatis, vascular inflow, and bile ducts

See **Figure 58.11a–d**.


Right hepatic duct
Cystic duct
Right hepatic artery
Common hepatic artery
(a)
Right hepatic artery
Hepatic artery
Common hepatic artery
(b)
Right portal vein
Portal vein
(c)
(d)

58.11a–d

CHOLECYSTECTOMY

This usually facilitates anatomic identification of the hilar structures. Division of the right portal vein into anterior and posterior sectoral branches occurs just deep to the medial gallbladder fossa. Note that the gallbladder should not be resected in a mesohepatectomy or trisectionectomy if a centrally placed tumor has a close margin in this area.

HILAR LYMPH NODES

Hilar lymph nodes, especially those surrounding the base of the celiac artery and the portal vein, are often enlarged. These nodes should be biopsied. Nodal involvement is very rare in HB, but it can be found in the other pediatric hepatic malignancies, especially HCC and rhabdoid tumors.

HEPATIC ARTERY

Arterial anatomy is notoriously variable, and identification of right and left arteries and any anatomic variants if present should be complete *before any arteries are ligated.* Bifurcation of the proper hepatic artery into right and left branches may be low (down by the gastroduodenal), intermediate, or high. There is usually one main artery to the right and left, but there may be two, even sometimes three, extrahepatic branches. Anatomic variant, or "replaced," right hepatic artery, will most often be found posterior and parallel to the common bile duct. Anatomic variant, or "replaced," left hepatic artery will usually course through the gastrohepatic ligament (~15% of cases).

PORTAL VEIN

Portal vein anatomy is less variable than arterial, but still all major vessels must be confirmed intact before the appropriate veins are ligated. The right portal vein quickly divides into two large anterior (segments 5 and 8) and posterior (segments 6 and 7) sectoral branches; this branch-point can be found just deep to the central aspect of the gallbladder bed. Branches may be divided individually if ligation of the common trunk would encroach upon the bifurcation. The left portal vein tends to be horizontal, has a longer extrahepatic course, and runs within the Rex fossa. Its branches to segments 2, 3, and 4 arise opposite each other. For an extended right hemihepatectomy (aka right trisectorectomy), the right portal vein is ligated and the branches to segment 4 are divided on the right or cephalad aspect of the left portal vein. For extended left hemihepatectomy (aka left trisectorectomy), the left portal vein and right anterior sectoral portal vein are divided.

PORTAL VEIN THROMBUS

Important: If portal tumor thrombus is encountered extending anywhere near the bifurcation, the surgeon should seriously consider whether a total hepatectomy with liver transplant might yield a better resection with less risk of local tumor relapse. Reconstruction of the portal vein after component dissection and resection of the vein with attached tumor thrombus has been reported but is considered extreme. The role of this type of resection and its results compared to liver transplantation has not been determined. In general, the decision to perform this type of operation should be done in consultation with a very experienced hepatobiliary surgeon in order to discuss the options.

BILE DUCTS

The bile ducts are undoubtedly the most inconstant feature and any anatomic uncertainty should be investigated with an intraoperative cholangiogram. The surgeon should not embark upon the liver parenchyma transection until the bile duct anatomy has been defined and dissected, and the bile duct to the remaining portion of liver has been preserved. Occasionally, it may not be clear what ducts can be spared or what reconstruction may be necessary until the parenchymal dissection is near completion. Bile duct reconstruction with an enteric loop is possible to one or more ducts in the case of extensive resection of large central tumors.

Parenchymal transection

PREPARE FOR PRINGLE MANEUVER

Before proceeding with parenchymal dissection it is important to confirm that the foramen of Winslow is patent and that it is possible to rapidly perform a Pringle maneuver to complete the vascular isolation using a vessel loop, umbilical tape, or atraumatic vascular clamp. Large tumors in segments 1, 4b, 5, or 6 may make this difficult. If so, the surgeon should simply note that inflow occlusion is not possible at this time and notify the anesthesia team of this fact.

DEMARCATION OF LINE OF DISSECTION

Once the arterial and portal inflow is divided, the ipsilateral liver demarcates and becomes dark. This color demarcation may be subtle and is best seen if the liver is not manipulated during the test clamp. This is the point at which the parenchymal traverse begins. Whatever the method employed to divide the liver, careful vigilance must be maintained in order not to wander off the plane of dissection that could potentially damage vital structures or bile ducts to segments of liver that are being preserved. Independent of the energy source or method chosen, the surgeon should be alert for large crossing vessels where suture ligation is often the most reliable method to prevent bleeding and risk of air embolism.

INTRAOPERATIVE ULTRASOUND FOR ANATOMIC GUIDANCE

Where tumor boundaries are indistinct, intraoperative US is very useful in anatomic location of tumor margins and the locations of variable hepatic venous drainage and crossing veins.

PARENCHYMAL DISSECTION DEVICES

The surgical approach to parenchymal transection can be made in a number of ways. Historically, this was done by finger fracture or clamp and crush techniques. Auxiliary tools that have been introduced to facilitate this process include the following:

- *Cavitron ultrasonic surgical aspirator (CUSA)* fragments and aspirates the hepatocytes along the dissection plane leaving the major blood vessels intact and visible. These can then be clipped or ligated and divided.
- *HydroJet*™ uses water energy to disperse hepatocytes from the underlying structures, again leaving the underlying vessels intact.
- *LigaSure*™ (Valleylab, Boulder, CO, USA) dissectors apply energy and pressure to divide tissue and seal blood vessels up to 7 mm in diameter.
- *Aquamantis*™ devices provide excellent hemostasis but have a larger zone of tissue destruction on either side of the line of resection. This can create occasional issues with interpreting margins of the pathologic specimen. When margins are not an issue, it is an excellent instrument.
- *GIA stapler*. Multiple firings of a GIA stapler will efficiently divide the liver with little bleeding. However, the width of the stapling device limits its utility in small children and infants, particularly with close margins.
- *Harmonic scalpel*□ (Ethicon Inc., Cincinnati, OH, USA) is perhaps the most popular in contemporary use. The advantages include better hemostasis and faster transection times. Care must be taken to avoid inadvertent thermal injury or unintended division of adjacent structures.

HEPATIC VEINS

Often divided prior to the onset of the parenchymal dissection, in select cases the hepatic veins may not be divided until completion of the parenchymal dissection if the hepatic venous margins are very close and challenging. The veins must be approached with caution and US or a CUSA dissector can be used very effectively to identify and/or skeletonize the veins and see them clearly before attempting to encircle and divide the veins to the affected side. Inadvertent uncontrolled tears in the major veins put the patient at risk for major bleeding and air embolus.

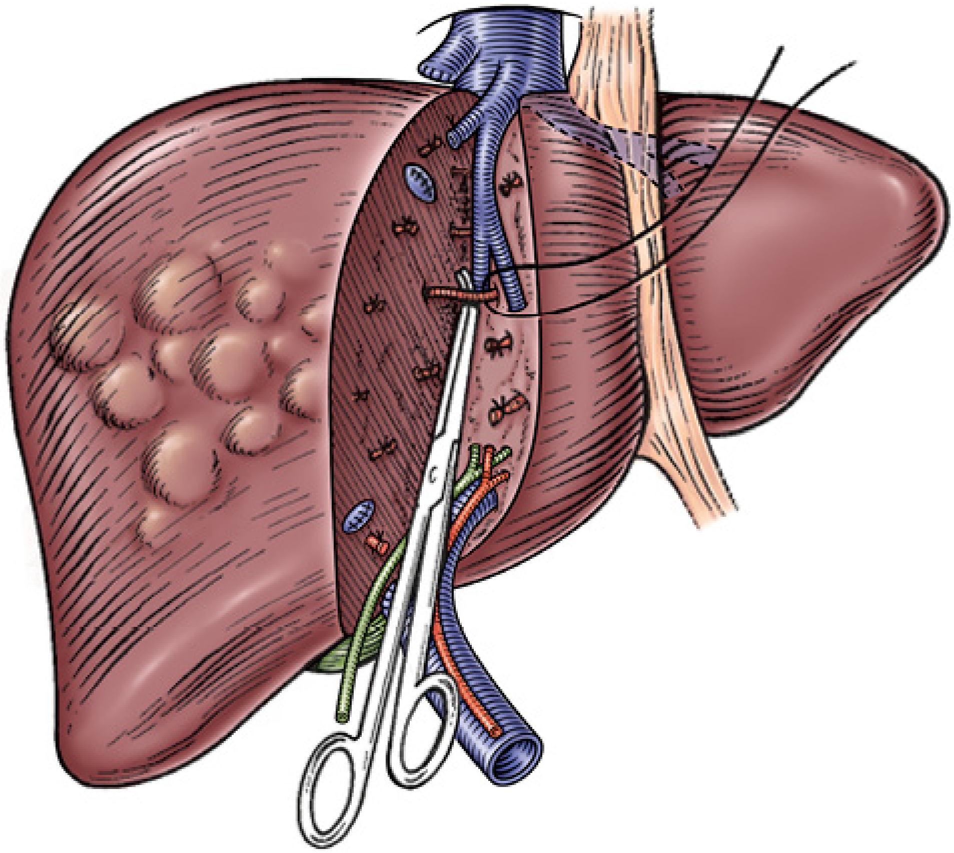
HEMOSTASIS

More than any other single technical aspect of the operation, strictly maintaining situational awareness and accurately defining anatomy will prevent any surgical catastrophe which would lead to serious hepatic dysfunction or death. Trial clamping of vessels before ligation and, if needed, use of the Doppler or intraoperative US is helpful. With vascular isolation maneuvers such as vena cava, hepatic vein, or portal triad (Pringle) clamping, short-duration intermittent clamping is often very helpful and is better than prolonged clamping. Patients are not usually anticoagulated when undergoing short periods of vascular isolation. Loss of control of a major hepatic vein is the most common disastrous intraoperative complication. During the vena cava dissection, which preceded the parenchymal dissection, plans will have been made for proximal and distal control of the cava, either with previously placed vessel loop/umbilical tapes or readily available preselected vascular clamps. Any sudden decrease in venous return accompanying a large venous opening will result in significant hypotension. Manual compression of the liver posteriorly can emergently reduce the bleeding with some restoration of venous return and allow the anesthesia team to catch up. Good communication between the teams while deciding when to fix the problem is critical. If necessary, formal vascular isolation with a Pringle maneuver and clamping of both infra and suprahepatic cavas will result in a dramatic reduction in bleeding and allow an expeditious but precise repair followed by improved venous return as the clamps come off (infrahepatic, followed by suprahepatic, followed by portal). Occasionally, there will be significant swelling of the residual segment 1 (caudate) if the venous drainage has been ligated without control of the portal branches supplying the segment, this may therefore require actual resection of the segment.

Removal of tumor

HEMOSTASIS AND RESIDUAL BILIARY DRAINAGE

Once the surgical specimen has been removed, hemostasis must be ensured. The vascular stumps should be checked for absence of bleeding. The cut surface of the liver must be inspected for bleeding and bile leaks, all of which must be oversewn (**Figure 58.12**). A completion cholangiogram may be performed to help confirm the absence of bile leak but also to check the integrity of the remaining biliary tree. Hemostatic agents such as Surgicel™, Hemospray™, Tisseal™, and Eviseal™ may be used on the raw surface at the surgeon's discretion but should not be depended upon to stop active bleeding.



58.12

MAINTAIN HEPATIC VENOUS OUTFLOW

If the ligamentous support to the remaining liver has been divided, it may advisable to suture the remaining liver to the remnant falciform ligament to prevent it from falling or rotating and kinking the venous outflow.

ASSESSMENT OF SURGICAL MARGINS

Gross surgical margins should always be negative. Increased margins (>1 cm) may be needed for resection upfront at diagnosis or for tumors that do not respond to chemotherapy (e.g. HCC). When there is any question, the surgeon should request an intraoperative pathologic assessment with inking and cutting of the specimen with both surgeon and pathologist assessing the gross margin. In any area where the gross margin is in doubt, if anatomically feasible, the surgeon should resect and submit additional margin for histologic analysis.

DRAINAGE

A surgical drain adjacent to the cut surface of the liver may be left in place at the end of the surgery before closing the abdomen. Drainage is not mandatory, but in many cases it is helpful to identify and control a bile leak in the early postoperative period. If a drain is placed, it is generally removed once a regular diet is resumed.

WOUND CLOSURE

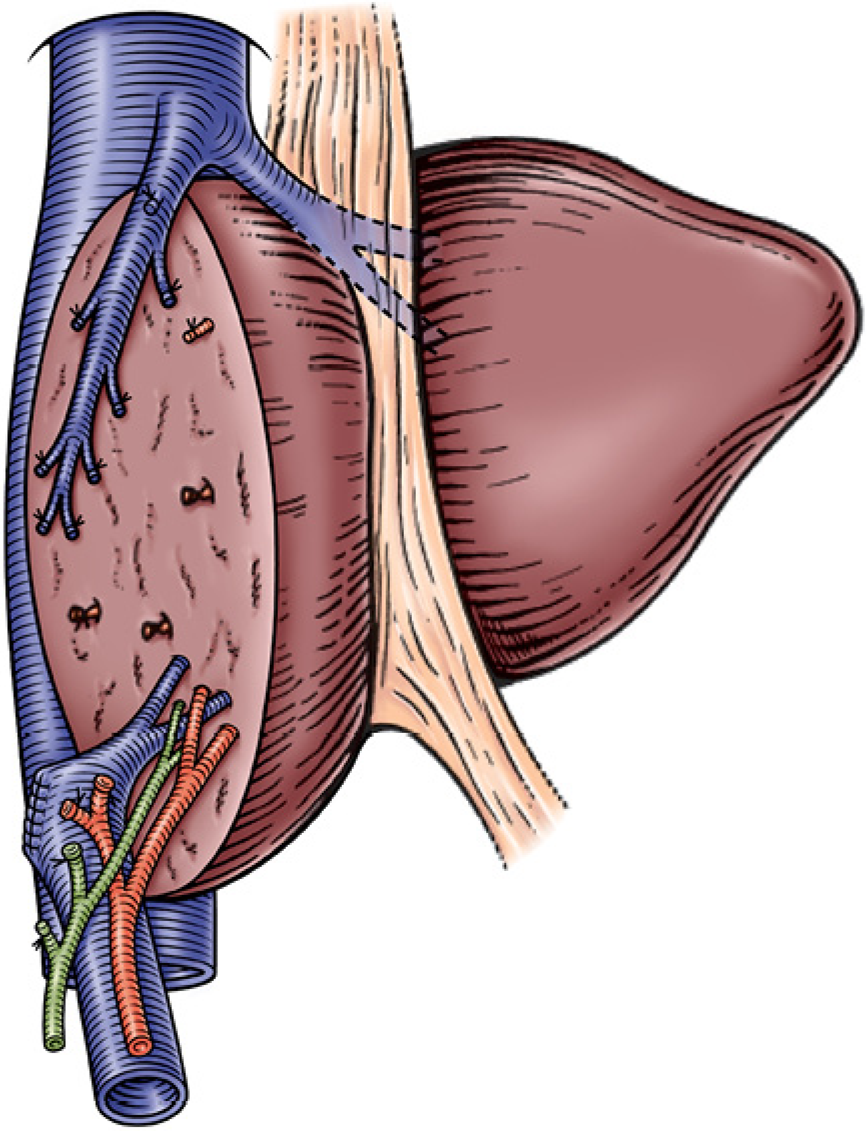
The abdominal wall is closed in anatomic layers with running, absorbable, monofilament suture.

SPECIAL CONSIDERATIONS FOR DIFFERENT TYPES OF LIVER RESECTION

Right-side tumors

RIGHT HEMIHEPATECTOMY (SEGMENTS 5, 6, 7, 8, AND SOMETIMES PART OF 1)

The abdominal incision should be carried quite lateral to facilitate exposure of the vena cava. The right adrenal gland and vein are preserved. It is important to consider the margins of the lesion from the portal bifurcation, vena cava, and especially the middle hepatic vein. This can ideally be done while the retractor is set up for the suprahepatic caval dissection. If inspection of the middle hepatic vein reveals an acceptable margin, the right hepatic vein may be dissected. Although this timing is a surgeon preference, having the ability to control the right hepatic vein before dividing the parenchyma decreases blood loss and the chance of an air embolism (**Figure 58.13**). Retrohepatic cava dissection will include division of short hepatic veins from segments 1, 6, and 7. If possible, a test clamp of right-sided inflow is performed and normal arterial and portal flow to segments 4a, 4b, 2, and 3 confirmed. If the result of the test clamp is satisfactory, the right portal bundle should be divided. During the parenchymal transection phase it is important to be constantly aware of the position of the tumor and the plane of dissection to prevent traveling to the left and injuring the middle hepatic vein. The caval portion of the dissection should have been completed but care should be taken to avoid transecting a hepatic vein not previously divided, or otherwise getting into the vena cava.



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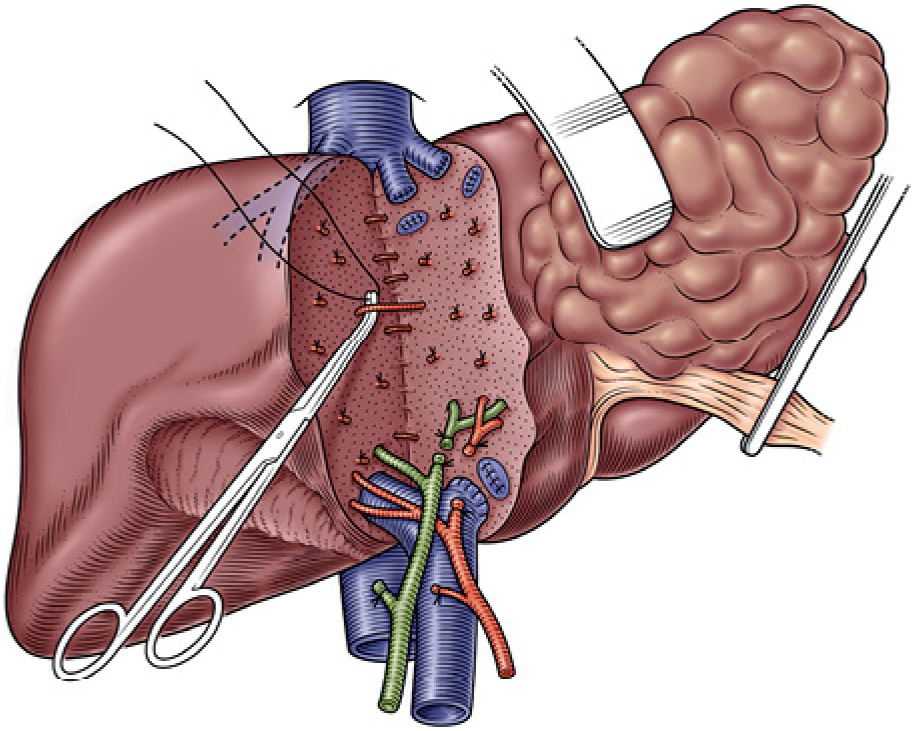
Left-side tumors

LEFT HEMIHEPATECTOMY (SEGMENTS 4A, 4B, 2, AND 3 WITH OR WITHOUT SEGMENT 1)

Exposure is optimally through an incision that is more central than that for a right-sided resection. Complete mobilization of the right lobe is not always necessary but mobilization of the left triangular ligament is required and provides exposure, with some additional dissection along the right side of the cava, to permit vascular control of the liver (**Figure 58.14**). A Kocher maneuver along with inflow control of the infrahepatic vena cava can be helpful if any reconstruction of the left and middle hepatic vein is required. Due to the variable anatomy of the middle vein, there is some ambiguity in terminology of resections. "Extended" left hepatectomies are those that include resection of the middle vein. The Brisbane classification focuses on segmental resections and not on the vessels involved. All of this may be of little interest to a surgeon trying to decide where to take the line of resection for a left-sided tumor. The large size of the right lobe as a liver remnant allows resection of the middle vein with little consequence if needed for a margin; if not needed for a margin, leaving the middle vein as the border of the resection is preferred.



58.14

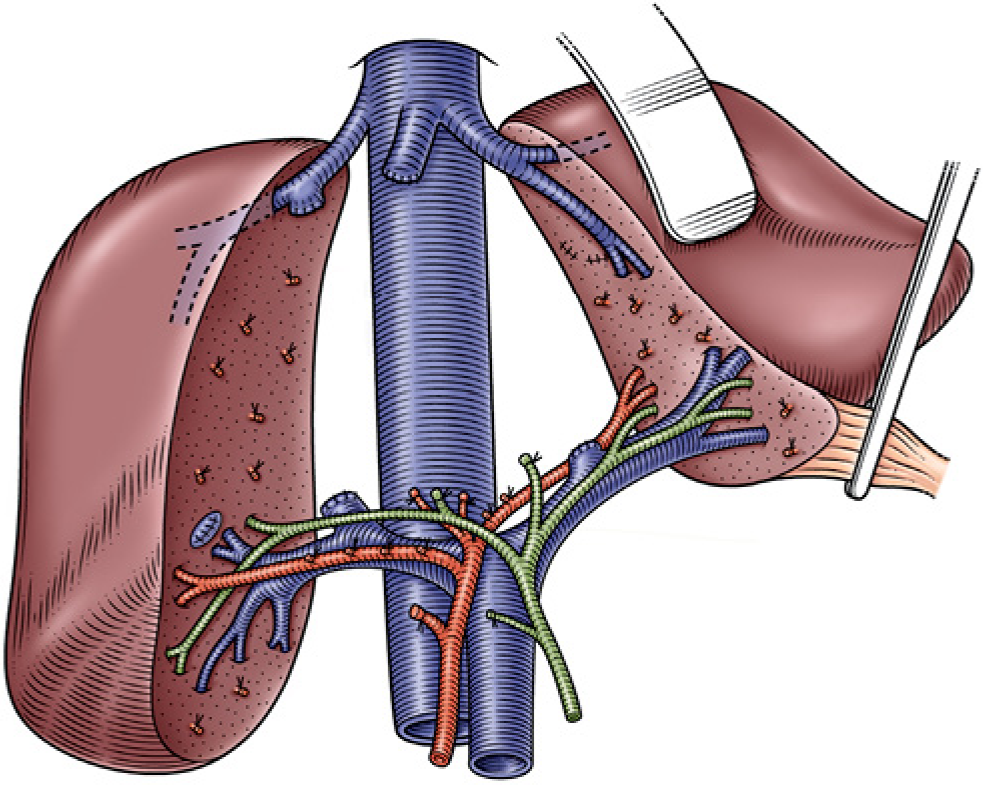
After sorting out the venous anatomy, the surgeon should check for a replaced left hepatic artery. Test clamping the vessel if it exists will demonstrate the area of the liver perfused. Usually an arterial branch from the common hepatic supplies segment 4 even when the "replaced" left is large. A separate left hepatic vein can be controlled early but should not be divided until after inflow control is achieved. If test clamps are satisfactory, the left portal inflow is ligated and divided, ensuring that the right portal inflow is intact. Whether or not a cholangiogram was performed, the surgeon would be well advised to realize that a segment 5 bile duct, or even the main right hepatic duct, may enter into the left system well into segment 4b, mandating a meticulous segmental biliary dissection. If the left vein joins the middle vein deep in the substance of segment 4a, and the intent is to save the middle vein, the left must be ligated and divided or, better, the resulting defect in the middle vein repaired under brief vascular control as needed. Resection of segment 1 requires ligation of multiple small hepatic veins draining directly to the vena cava, which potentially places segment 6 and 7 veins at risk. Margins permitting, an Endo GIA™ stapler can be fired, splitting segment 1 above the cava, preserving the right-sided hepatic veins, and making quick work of this pesky area. Blood loss from the hepatic veins is usually less in left-sided resections than in right-sided ones but the anterior location of the vein orifices makes air embolization more likely. Carefully clamping and suturing of the veins or stapling them without opening them first best prevents this problem.

LEFT LATERAL SECTORECTOMY (SEGMENTS 2 AND 3)

The ease of this operation should not lead the surgeon to skip important steps including consideration of intraoperative US to develop a clear mental map of the 3D anatomy. If a replaced left hepatic artery exists, it will need to be identified and divided. Widely opening the gastrohepatic ligament and staying above the nodes in the porta, the surgeon can elevate the sector and achieve excellent exposure. If there is a separate left hepatic vein, it may be dissected and circled with a vessel loop. An umbilical tape passed from along the medial aspect of the left hepatic vein, into the lesser sac, and then inferiorly to the Rex fissure allows one to completely encircle the line along which the resection is to be performed. In order to avoid injury to segment 4 ducts, the left portal vein or arteries to the remnant liver, the individual bundles to segment 3 and then segment 2 should be taken individually. They are found along the ligamentum teres, retracting segments 2 and 3 to the left and superiorly opening up the Rex fissure. If this runs the risk of compromising the margin, the surgeon should consider a formal left hemihepatectomy.

Central liver lesions

A central resection is more complex than those limited to the right or left hemilivers since this requires the surgeon to perform a portal dissection on both the left and the right main portal branches and thus put the entire liver at some risk. Nonetheless, mesohepatectomy leaves the child with significantly more residual liver segments and thus the physiologic impact is less than a trisectorectomy (**Figure 58.15**). The necessity for significant dissection before any test clamping can be done requires the surgeon to have a detailed understanding of the anatomy of the liver and the position and size of the tumor, and to commit to the resection early. Before making this commitment, a plan for handling the middle hepatic vein, and its common wall (if present) with the left hepatic vein must be made so that any resulting defect may be closed without obstructing the outflow of the left lateral sector. Obtaining vascular control of the vena cava is an important precaution to take for any mesohepatectomy. There will be two large parenchymal transections with the resulting raw area and issues with hemostasis and bile leaks. Margins of resection on the left are the Rex fissure, then vertically to the junction of the left and middle hepatic veins. On the right, the line of resection extends from near the infundibulum of the gallbladder, laterally to just anterior to the right hepatic vein. In the hilum of the liver, the resection line is nearly horizontal and just above the portal vein. Superiorly, the line extends from the junction of the left and middle hepatic vein to just anterior to the right hepatic vein.



58.15

CAUDATE LOBE RESECTION

The surgeon will need to mobilize both the left and right hemilivers. It is tempting to take the hepatic vein branches to segment 1 during this mobilization, but this is a mistake as swelling of the segment makes rupture almost inevitable with the amount of pulling and tugging needed during the parenchymal dissection. After rotating the right hemiliver up in the air, the surgeon, often crouched in an awkward position, must free the porta hepatis from the segment by taking the posterior branches while carefully preserving those to the anterior sector. Working alternately from the right and the left sides is often helpful, especially as the left liver is considerably more mobile than the right. After the inflow is controlled, sequential ligation of the direct hepatic vein branches frees the cava from segment 1. Most of these may be accessed from the patient's right, but some will be better seen through the opened gastrohepatic ligament on the left. After completing this dissection, the parenchymal transection is begun inferiorly and carried superiorly to the apex of the resection, taking care not to stray anteriorly or laterally into the right posterior sector. The superior point of resection is a narrow wedge between the cava and the middle hepatic vein.

Extended resections including left trisectorectomy (segments 1, 2, 3, 4, 5, 8) or right trisectorectomy (segments 4a, 4b, 5, 6, 7, 8)

The challenge of these operations is the small-for-size remnant liver volume and the need for a high level of technical precision to avoid any compromise to this remnant since the patient will already be marginally compensated from a metabolic standpoint. Most of these extended liver resections are done for very large tumors. Extensive multifocal tumors are sometimes better managed by complete hepatectomy and liver transplantation. Occasionally, smaller tumors with macrovascular invasion are so tightly adherent to the cava, or are situated at the confluence of the hepatic veins, as to necessitate vascular resection and reconstruction to achieve complete tumor resection. These situations also should prompt consideration of a liver transplant. A variety of issues inform this decision and careful, collaborative treatment planning should be done before embarking on any of these resections.

The liver remnant after a *left trisectorectomy* is the right posterior sector (segments 6 and 7). The preserved portal bundle is the right posterior sector bundle with venous outflow based on preserving the right hepatic vein. The anatomy and technical aspects are largely covered above with a few key, and important, issues for the surgeon to consider. The right lobe should not be mobilized off the vena cava because important auxiliary venous drainage of segments 6 and 7 should be preserved. Caudate short hepatic veins should be approached first inferiorly and then from the left, although leaving a portion, or all of segment 1 in place is preferred if margins allow this luxury. The gallbladder is usually taken en bloc with the specimen. The left portal bundle can be taken after the go/no go decision is made. Depending on the margin, dissection of the right portal bundle can be within the Glissonian sheath or outside it. If the dissection is done within the sheath, care must be exercised not to make the hepatic duct ischemic. Isolation and test clamping of the anterior portal bundle allows confirmation that the liver remnant has adequate arterial and portal venous flow. After confirming this and taking the anterior sectoral bundle, the middle and left hepatic vein may be sacrificed. The parenchymal transection will be on a sagittal plane just above the right hepatic vein.

The liver remnant after a *right trisectorectomy* is the left lateral sector, segments 2 and 3. The preserved portal bundle is the left portal bundle minus the medial branches to segments 4a and 4b. The venous outflow is the left hepatic vein. The right lobe is extensively mobilized off the vena cava, with or without segment 1, and the right portal bundle is divided after making the go/no go decision. The right hepatic vein may then be controlled and divided. Dissection along the superior border of the left portal bundle and reflecting that bundle inferiorly proceeds to the point where it turns vertically in the Rex fissure. The parenchymal dissection is the same as that described for mesohepatectomy, i.e. ligating the medial arterial, portal, and biliary branches to segments 4b and 4a.

Extreme resections and vascular reconstruction

Resection and reconstruction of major vasculature to the remnant liver and/or the vena cava is sometimes an alternative to complete hepatectomy and liver transplantation. With the success of liver transplantation and excellent oncologic results, even in children with controllable metastatic disease, the number of these operations is relatively small. The surgeon should be able to predict the need for any of these techniques during the preoperative planning phase and should not undertake these “on the fly” based on intraoperative findings. The operations all involve some degree of warm ischemic time to the remnant liver and they should therefore be undertaken only in consultation and collaboration with a specialized liver surgeon. Materials useful in reconstruction during these cases include direct repair for small defects where the repair does not compromise the diameter of the vein. Grafts to replace or patch the vena cava, suprahepatic vein, or portal can include cadaveric donor veins, polytetrafluoroethylene (PTFE), or autologous internal jugular. It is key to design an approach to each potential vascular reconstruction that minimizes ischemic time to the remnant liver. If there is any chance that the total ischemic time might be prolonged over 30 minutes, an alternative plan should be sought. Tolerance of the liver to ischemia is dramatically improved by cooling. This is most effectively done by perfusing the liver with ice-cold isotonic fluid combined with surface cooling, but the cold fluid must not be infused into the heart as major arrhythmias or cardiac arrest can occur. This technique, commonly used by transplant teams, is only needed during extreme resections with major vascular reconstructions and should not be used without special training and considerable experience.

POSTOPERATIVE CARE

Following major liver resection patients are admitted to the pediatric intensive care unit for continuous hemodynamic monitoring.

Intraoperative resuscitation is continued to:

- Maintain or restore core temperature to >36.5°C
- Maintain or restore urine output of 0.5–1 mL/kg/hour
- Hematocrit >22%
- Platelet count >80 000
- International normalization ratio (INR) <1.8

Laboratory interrogation for hepatic insufficiency will include serial measurements of lactate, glucose, PT/INR, fibrinogen, and acid–base balance. Over-resuscitation is avoided and a CVP <8 is preferred, provided other indicators of perfusion are appropriate. Closed suction drainage is monitored for volume and quality of output. A clear liquid diet is started once the signs of postoperative ileus resolve. The drain may be kept in place to monitor for bile leak until the patient is tolerating a full diet. Perioperative antibiotics are usually discontinued after 24 hours. Beginning within 24 hours of resection, hypophosphatemia, hypokalemia, and hypomagnesemia are common and these electrolytes should be replaced appropriately.

COMPLICATIONS

A number of possible complications can follow liver resection.

- *Hemorrhage.* This should be suspected in a patient with poor perfusion, labile hemodynamics, inappropriate response to transfusion, or failure to improve with resuscitation. Drainage may be bloody, but, when plugged or loculated, the drain may not be a reliable indicator of bleeding. When in doubt, the surgeon should maintain a low threshold for operative re-exploration and accept the possibility of a negative exploration rather than delay the operative control of significant bleeding.
- *Hepatic dysfunction.* Liver failure after liver resection is usually due to ischemia, either from temporary clamping of the vascular structures to limit blood loss during the resection, or from inadvertent damage or ligation of vessels to the remnant liver, or from too-small-for-size remnant liver. The degree of hepatic dysfunction can be monitored by serial lactate, glucose, PT/INR, and fibrinogen levels. Hypoglycemia is monitored and treated with 10% dextrose, elevated PT/INR is treated with FFP and sometimes exogenous vitamin K, and fibrinogen level <100 mg/dL is treated with cryoprecipitate. Transient elevation in the liver AST/ALT levels and bilirubin levels are common but more prolonged elevation can signal hepatic arterial insufficiency. If the acute vascular insufficiency is severe, dysfunction of the remnant liver can lead to encephalopathy, coagulopathy, hypoglycemia, and death within several days. Prevention is key since, once this has happened, repairs involve further ischemia reperfusion injury and often do not result in improvement in the liver's function in the critical window of time left. Emergent rescue liver transplants have been done but the survival after these is not good.
- *Hepatic congestion from obstructed venous outflow.* Obstruction of the hepatic veins will create a Budd–Chiari physiology with venous congestion in the intestines, ascites and significant liver dysfunction, and may be fatal. Venous obstruction may cause swelling of the liver remnant and venous hypertension will increase the risk of bleeding from the cut surface. If any question exists about the ability to achieve a complete

resection without compromising the venous drainage of the remnant liver, the planned resection needs to be modified. Always remember that results of total hepatectomy and liver transplantation for HB are excellent. Thus, early decision before bridges are burned is key. Experienced surgeons have been successful in doing "extreme" resections with vascular repair of the hepatic veins or cava, but such an approach needs to be very well thought out and is likely to increase the risk of complications. Intraoperative US to define the anatomy of the hepatic veins prior to parenchymal dissection is key to avoiding this complication.

- *Bile leaks.* These can have serious consequences including portal hypertension and suboptimal liver regeneration. Most are self-limiting and can be treated with appropriate controlled drainage. Persistent leaks may be associated with obstruction of the remnant biliary system and, at worst, may require internal drainage of the leak with a Roux-en-Y jejunal loop.
- *Renal dysfunction.* This is unusual and can be related to prolonged obstruction of the renal veins. Postoperative renal dysfunction after a vena cava reconstruction should prompt investigation of the reconstruction.
- *Delayed gastric emptying.* This may result from extensive node dissection in the region of the lesser curve.
- *Portal hypertension.* Immediate postoperative venous congestion of the bowel may result from encroachment on the remaining portal vein from a misplaced ligature or kinking/twisting of an excessively mobile remnant or thrombus. If the postoperative US suggests a technical error, it should be corrected. Late-onset portal hypertension may rarely result from biliary cirrhosis or ischemic fibrosis.
- *Pleural effusions or a pneumothorax.* These may occasionally complicate a right hemihepatectomy and can be handled by aspiration or a thoracostomy tube. Persistent pleural drainage suggests a subdiaphragmatic bile collection, hematoma, or injury to the lymphatics forming the thoracic duct at the caval hiatus.

OUTCOME

Hepatic regeneration is remarkably fast and most patients will have a normal liver volume within several months of liver resection. Overall outcome is related to the patient's underlying diagnosis. Successful resection is generally curative for benign disease, while resection remains the cornerstone of therapy for primary hepatic malignancies.

VIDEOS

OPS_8e_video_58.1 Incision for major liver resection
OPS_8e_video_58.2 Inferior vena cava exposure
OPS_8e_video_58.3 Non-anatomic liver wedge resection
OPS_8e_video_58.4 Right hemi-hepatectomy
OPS_8e_video_58.5 Right trisectorectomy
OPS_8e_video_58.6 Gross section

FURTHER READING

Aronson DC, Meyers RL. Malignant tumors of the liver in children. *Semin Pediatr Surg* 2016; 25: 265–75.

Brock PR, Maibach R, Childs M, et al. Sodium thiosulfate for protection from cisplatin-induced hearing loss. *N Engl J Med* 2018; 25: 2376–85.

Cornet M, DeLambert B, Pariente D, et al. Rhabdoid tumor of the liver: Report of pediatric cases treated at a single institute. *J Pediatr Surg* 2018; 53: 567–71.

Czauderna P, Haeberle B, Hiyama E, et al. The Children's Hepatic tumors International Collaboration (CHIC): Novel global rare tumor database yields new prognostic factors in hepatoblastoma. *Eur J Cancer* 2016; 52: 92–101.

Langham MR, Furman WL, Fernandez-Pineda I. Current management of neonatal liver tumors. *Curr Pediatr Rev* 2015; 11: 195–204.

Lopez-Terrada D, Alaggio R, De Davila MT, et al. Towards an international pediatric liver tumor consensus classification: Proceedings of the Los Angeles COG International Pathology Pediatric Liver Tumors Symposium. *Mod Pathol* 2014; 26: 19–28.

Meyers RL, Tiao G, de Ville de Goyet, et al. Hepatoblastoma state of the art: PRETEXT, surgical resection guidelines and the role of liver transplantation. *Curr Opin Pediatr* 2014; 26: 29–36.

Meyers RL, Maibach R, Hiyama E, et al. Risk stratified staging in paediatric hepatoblastoma: A unified analysis from the Children's Hepatic tumor International Collaboration (CHIC). *Lancet Oncol* 2017; 18: 122–31.

Murawski M, Weeda VB, Maibach R, et al. Hepatocellular carcinoma in children: Does modified platinum- and Doxorubicin-based chemotherapy increase tumor resectability and change outcome: Lessons learned from the SIOPEL 2 and 3 studies. *J Clin Oncol* 2016; 34: 1050–56.

Strasburg SM. Nomenclature of hepatic anatomy and resections: A review of the Brisbane 2000 system. *J Hepatobiliary Pancreat Surg* 2015; 12(5): 351–5 PMID 16258801.

Towbin AJ, Meyers RL, Woodley H, et al. 2017 PRETEXT: Radiologic staging system for primary hepatic malignancies of childhood revised for the Paediatric Hepatic International Tumour Trial (PHITT). *Pediatr Radiol* 2018; 48: 536–54.

59

Wilms' tumor

KEVIN CAO, DENIS A. COZZI, AND PETER M. CUCKOW

HISTORY

Nephroblastoma was first described in a series by Max Wilms, a German surgeon and pathologist with an interest in renal pathology, in 1899. The first successful nephrectomy for such a tumor in a child was carried out by British surgeon Thomas Richard Jessop in 1877. Radiotherapy was introduced later as adjunctive therapy in 1916 by Alfred Friedlander in Cincinnati. In 1956, Sydney Farber introduced the chemotherapeutic agent actinomycin into their treatment regimen, ushering in modern chemotherapeutic protocols for this disease.

Modern treatment protocols that combine chemotherapy, surgery, and selective radiotherapy by a multidisciplinary team of surgeons, oncologists, radiologists, pathologists, pediatricians, and specialist nurses, now achieve >85% long-term survival rates. Much of the current focus in Wilms' management is data collection and assessment of the prognostic value of specific mutations, biobanking excisions to enable cross-national collaboration in evaluating genomic drivers, and collecting collateral tissue such as blood and urine in order to discover and test new biomarkers.

PRINCIPLES AND JUSTIFICATION

Nephroblastoma (Wilms' tumor (WT)) is a renal malignancy thought to be derived from embryonic renal tissue, often with metanephric blastema, incomplete tubules, and stromal tissue. In 90% of cases it presents as a painless abdominal mass in an otherwise well child and affects one child per 10 000 worldwide annually, with slight female preponderance. While rare overall, it is the most frequent primary renal tumor in childhood, representing ~7% of all pediatric cancers.

- 80% of cases are diagnosed <5 years of age, with peak incidence between 1 and 3 years.
- ~ 6% are bilateral.

Currently, there are two strategies for the treatment of WT. The North American National Wilms' Tumor Study Group (NWTSG), which merged into the Children's Oncology Group (COG) in 2002, advocates upfront nephrectomy followed by chemotherapy, while the Société Internationale d'Oncologie Pédiatrique (SIOP) advocate pre-nephrectomy chemotherapy to shrink the tumor before surgery. Chemotherapeutic agents vary based on tumor staging in both protocols but use of dactinomycin and vincristine is present in most regimes. In the UK, the 2016 updated UMBRELLA SIOP-RTSG protocol is used, with pre-chemotherapy biopsy foregone in virtually all cases due to the risk of upstaging of tumors. Secondary deposits, caval tumor, bilateral disease, or tumor in a solitary kidney are indications for chemotherapy and delayed surgery.

PREOPERATIVE ASSESSMENT

Usual workup should include full blood count, liver and renal biochemistry, and coagulation studies (von Willebrand's disease is a rare association with WT). Genetic analysis should include mutations analysis such as 11p13 (WAGR syndrome), 11p15 (Beckwith–Wiedemann syndrome), and WT1 (Denys–Drash syndrome).

Radiology

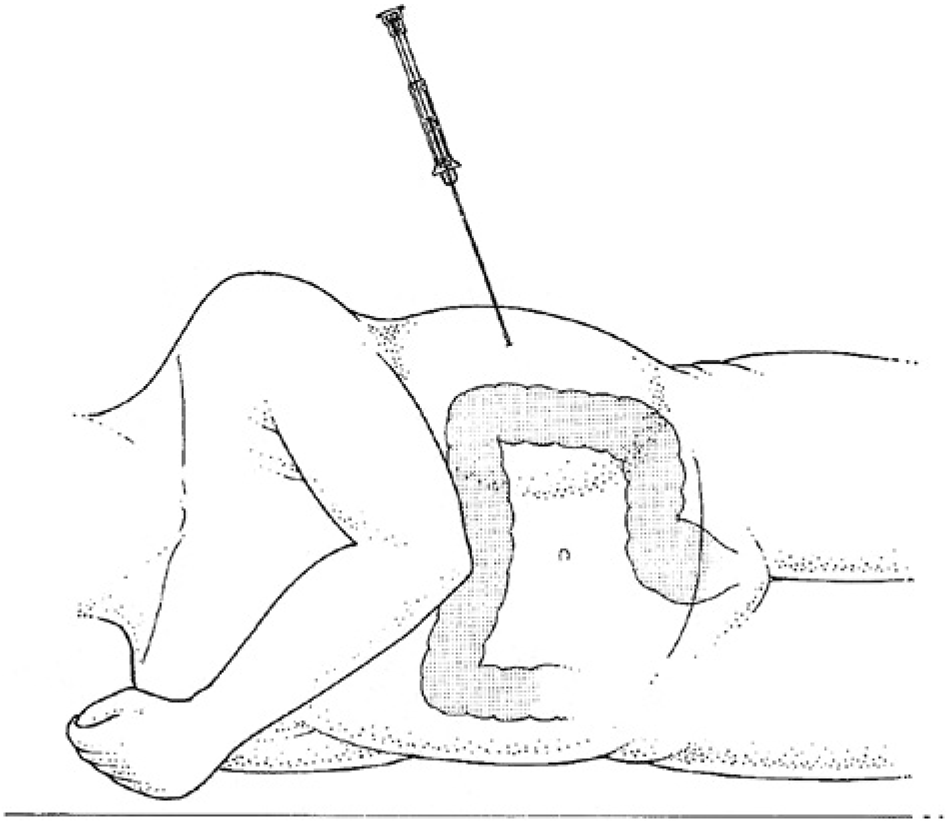
Ultrasound (US) is usually the first imaging modality to be used and will demonstrate a solid tumor arising from the kidney, and utilizing Doppler assessment can evaluate for renal vein or vena cava involvement. US can also identify liver or abdominal metastases and survey the contralateral kidney, where smaller deposits may denote bilateral WT.

Chest radiography (anterior and lateral) is still used in European trials to identify pulmonary metastatic disease, but computed tomography (CT) scans are more sensitive and provide more complete information about the lungs, kidneys, and state of local invasion. In nephron-sparing surgery (NSS), the creation of 3D images that combine angiography, venography, and urography may be particularly useful. Although in the past exploration of the contralateral kidney was a mandatory part of the operation, this has now been replaced by CT scan. Nevertheless, it is still the surgeon's role to determinate inoperability at exploration.

Biopsy of the tumor

Biopsy in the UK is no longer favored due to the risk of upstaging of cystic lesions in the case of wedge-type biopsies. The SIOP protocol used across Europe generally advises histologic diagnosis based on the post-chemotherapy nephrectomy specimen. However, there are certain occasions where a percutaneous biopsy can be helpful, such as in stage 4 disseminated diseases, where it may guide oncologic treatment, in children over 10 years of age where non-Wilms' lesions are more common (e.g. rhabdoid tumor, renal cell carcinoma), and in those cases where imaging are strongly suggestive of an alternative diagnosis.

With the patient in the lateral oblique position, Trucut™ biopsy is best performed under US guidance (**Figure 59.1**). Bleeding can occur if the tumor is particularly cystic, however tumor seeding into the track is rare. Minimally invasive laparoscopy has the potential of a more active role, particularly in assessing and taking biopsies of intra-abdominal tumor spread.

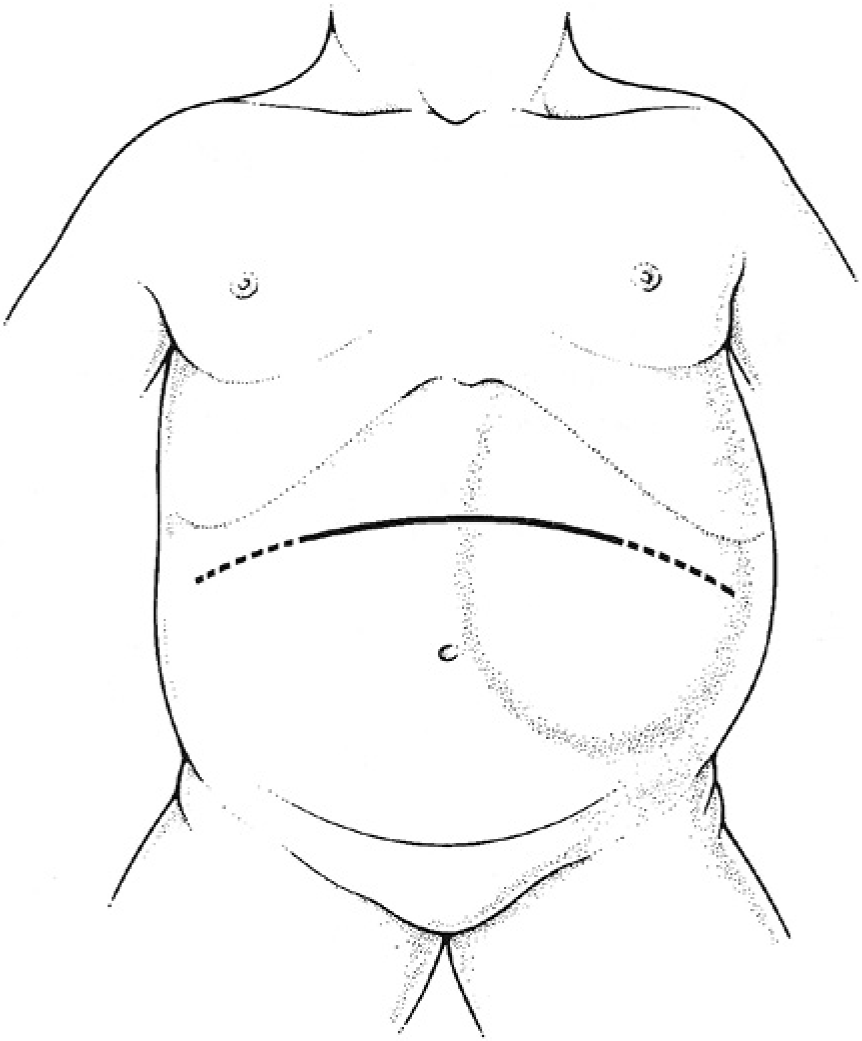


59.1

OPERATIONS

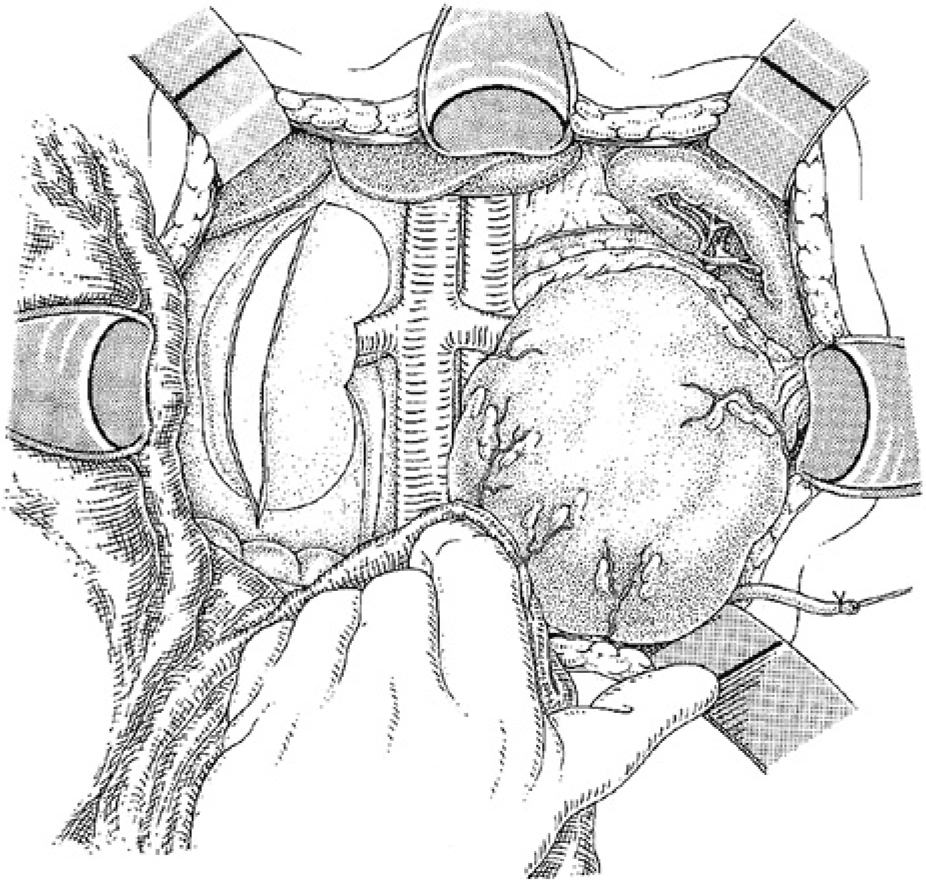
Nephrectomy

Transverse upper abdominal incision starting from on the side of the tumor, extending across the contralateral rectus muscle provides complete exposure of the kidney and access to its pedicle (**Figure 59.2**). A longitudinal incision extending from the xiphoid to below the umbilicus may be more suitable in older patients.



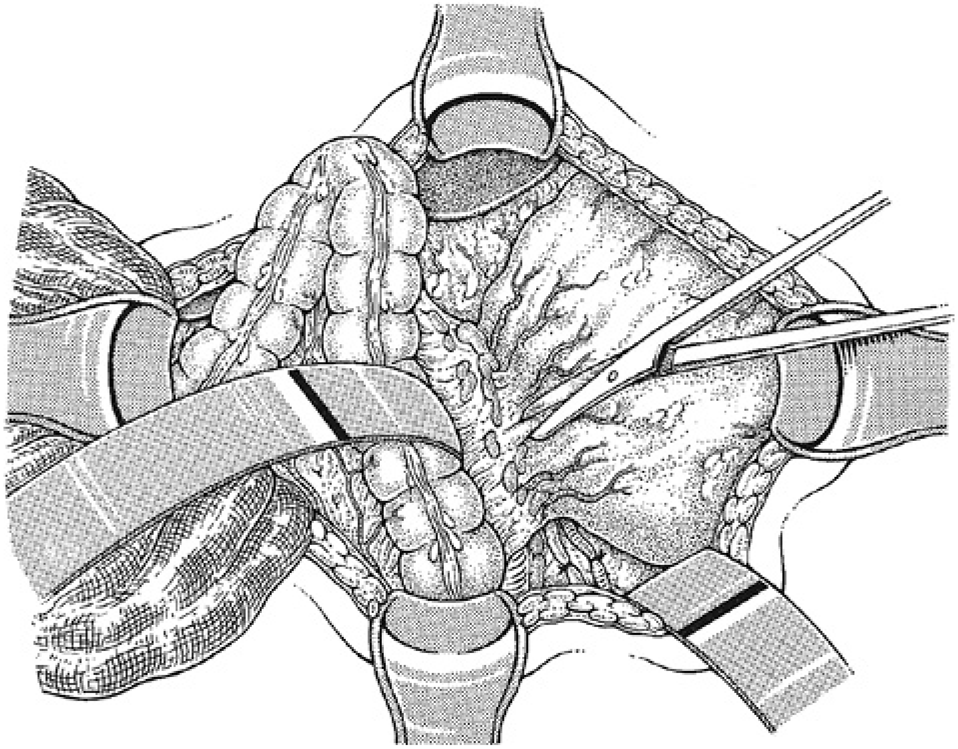
59.2

Traditionally, the contralateral kidney may be mobilized and examined in both anterior and posterior surfaces to detect possible bilateral disease (**Figure 59.3**). If a CT scan has been performed, the visual and manual inspection of the contralateral kidney is at the discretion of the surgeon. If unsuspected bilateral disease is detected, the main tumor and contralateral lesion are biopsied with no further excisional surgery. The patient is subjected to chemotherapy with subsequent NSS.



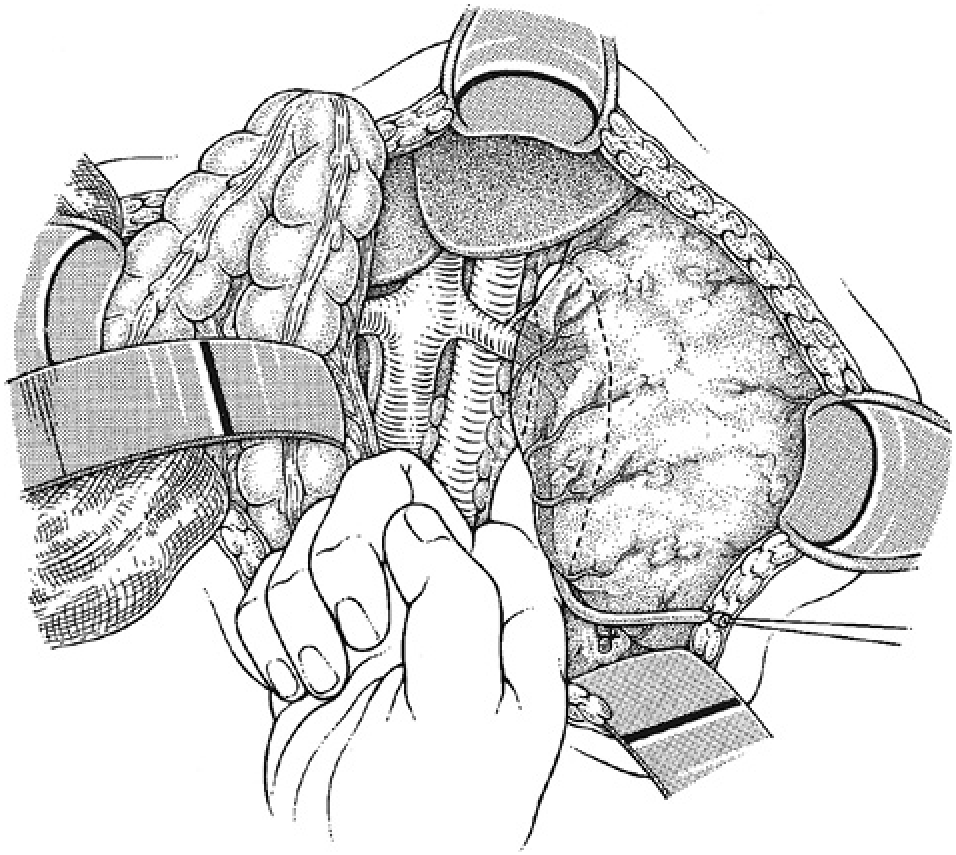
59.3

The colon with its mesentery is dissected carefully from the anterior surface of the tumor after first dividing the lateral peritoneal attachment (**Figure 59.4**).



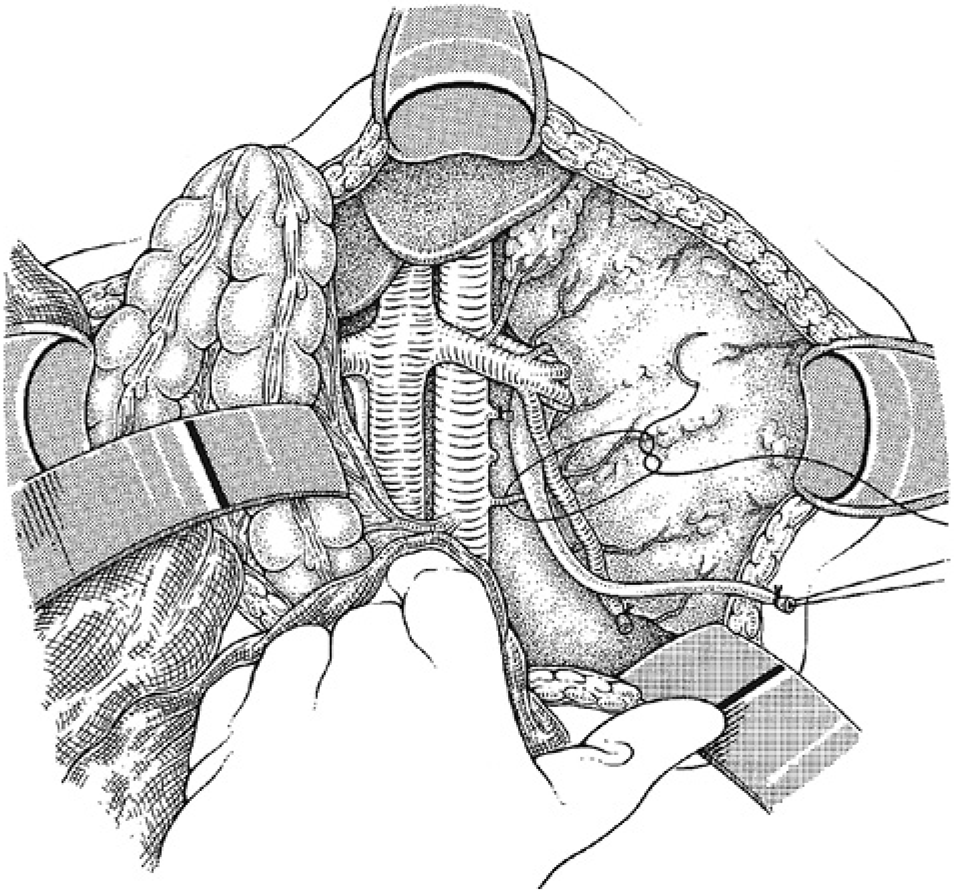
59.4

The ureter and gonadal vessels are ligated well below the kidney and may now be transected (**Figure 59.5**). The renal vessels cross in front of the ureter and renal pelvis, so this is used as a guide to find them as dissection continues cranially. If possible, the tumor is not mobilized at this stage, although large tumors which cross the midline can distort the anatomy. Lateral mobilization of the kidney can create more medial space in these cases and may require a further lateral extension of the incision.



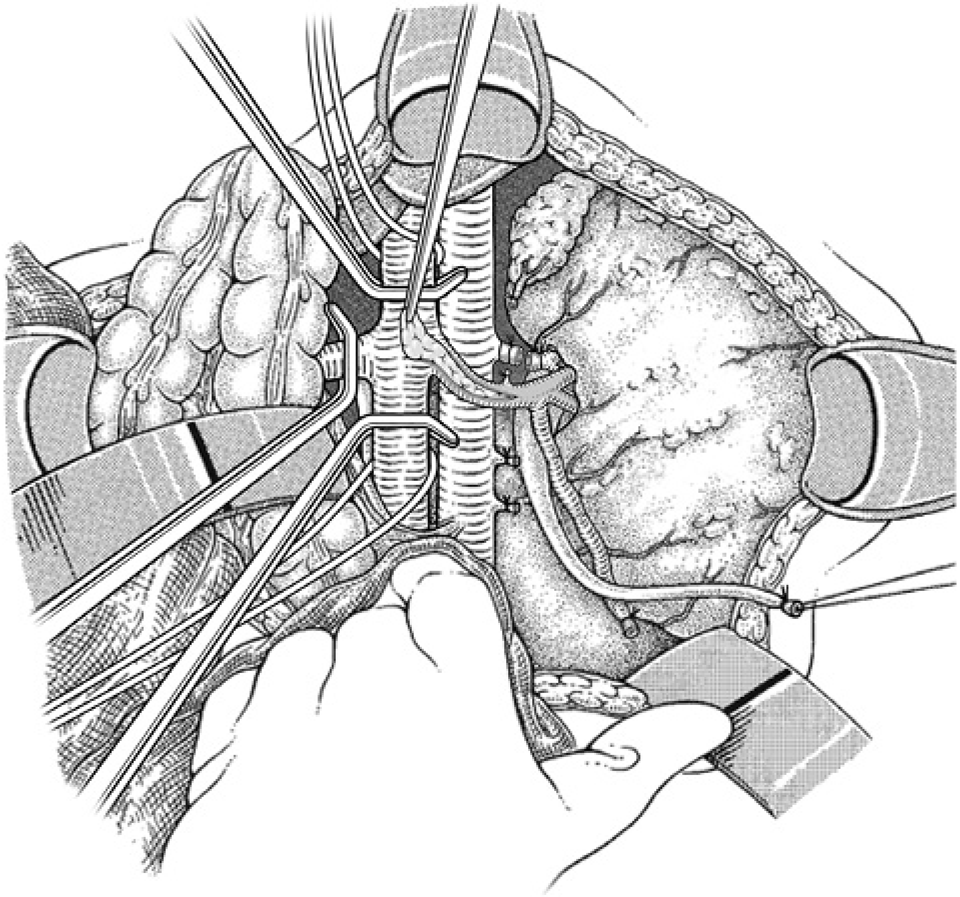
59.5

Dissection begins caudally, sweeping adventitial tissue and lymph nodes laterally off the great vessels, if necessary, dividing it between ligatures (**Figure 59.6**). The renal vein and its branches are gently exposed. Careful palpation of the vein may detect venous extension of the tumor, although this should have been noted on preoperative investigations. Early mobilization of the inferior vena cava and renal vein may be required to prevent embolization of the tumor into the inferior vena cava, heart, and pulmonary artery. On the right side, the second part of the duodenum is encountered during this dissection and is reflected medially off the vessels.



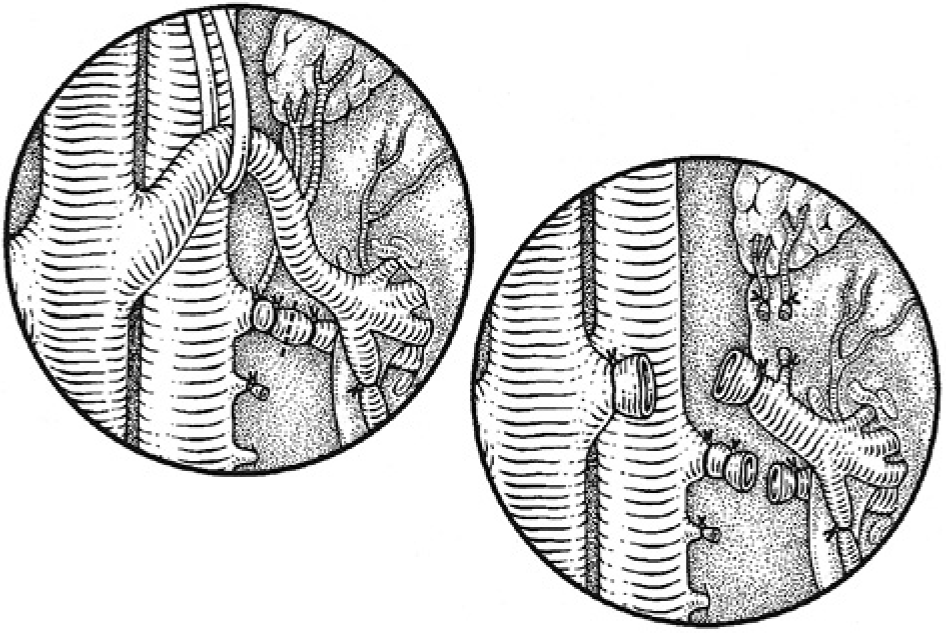
59.6

If preoperative investigation has discovered vena caval involvement with malignant thrombus, this must be extracted via a venotomy. Inferior vena cava control can be obtained by placing slings above and below the level of tumor thrombus and around both renal veins (**Figure 59.7**). If access is difficult, a right-sided approach after Kocherization of the duodenum may be necessary. A probe may be needed to dissect the thrombus from intimal attachments.



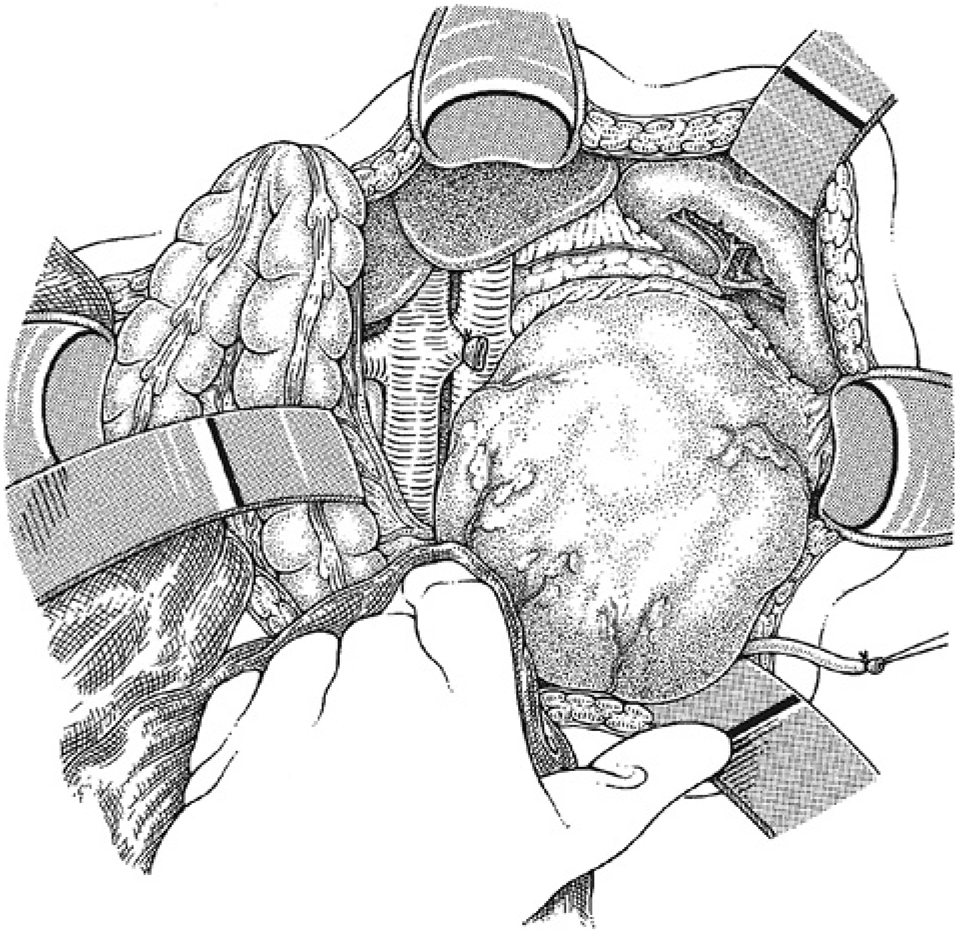
59.7

The renal vein is gently mobilized and elevated with a vascular sling to expose the renal artery, which is typically situated behind the upper border of the renal vein (**Figure 59.8**). It is preferable to ligate and divide the renal artery first so as to prevent swelling of the kidney with arterial blood. Due to the size of the tumor, this sequence of ligation is not always possible.



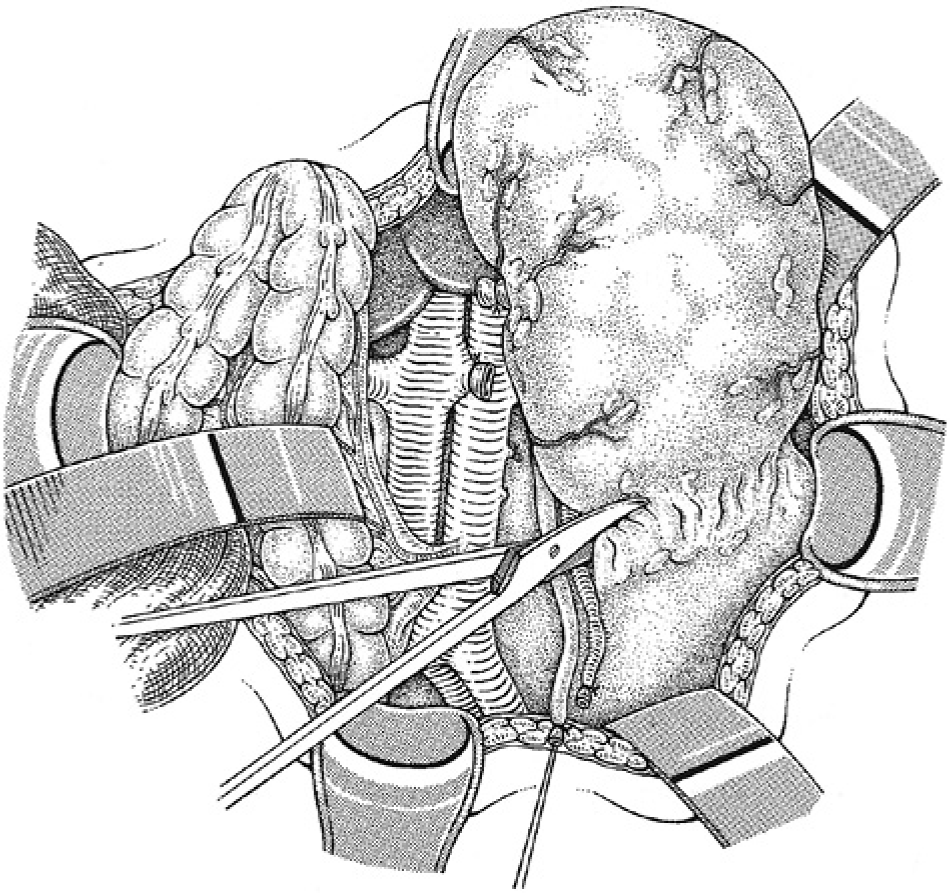
59.8

The posterior aspect of the kidney is partially mobilized by blunt dissection. The superior dissection is hazardous on the left side, and damage to the spleen and tail of the pancreas must be avoided. On the right side, the tumor may be stuck to the under surface of the liver, but rarely infiltrates it. Care is taken in separating them to avoid breaching the liver capsule (**Figure 59.9**). The adrenal gland is removed only if the tumor is in the upper pole of the kidney. In this circumstance, the adrenal vessels should be carefully controlled with ligatures.



59.9

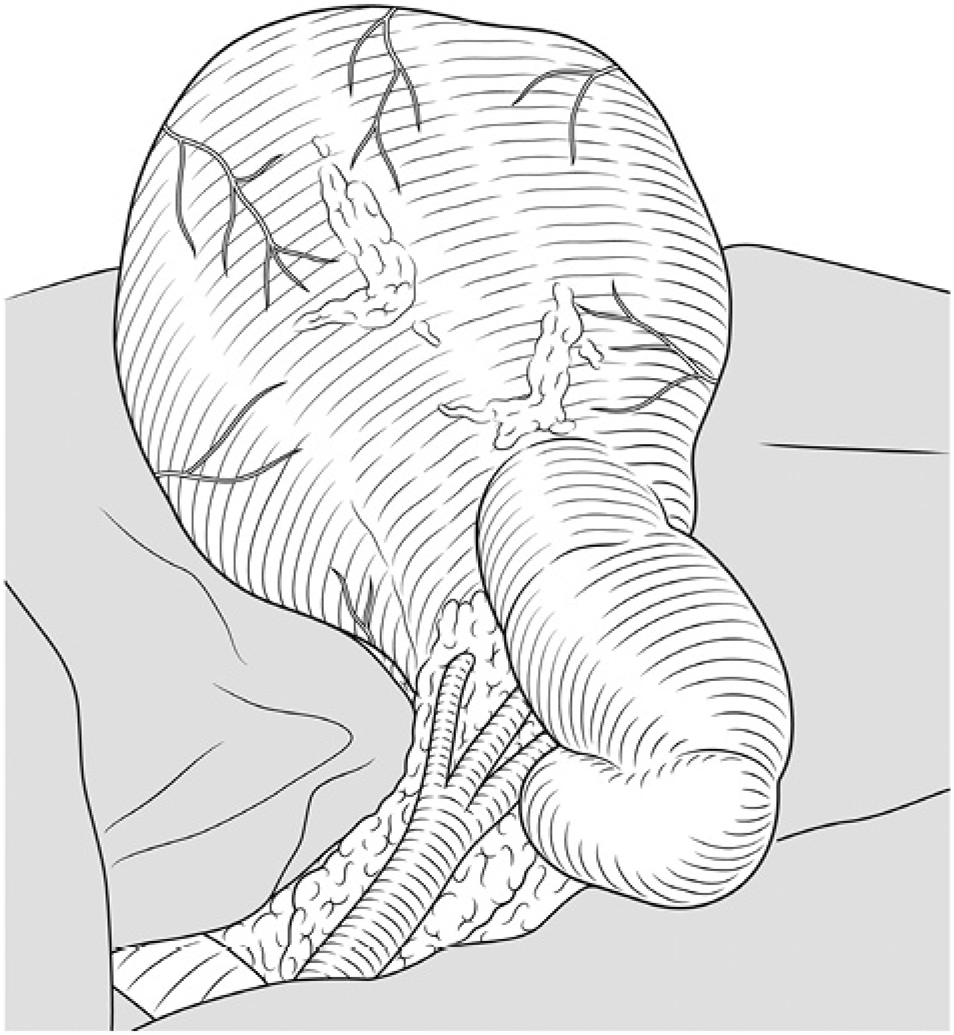
The kidney within Gerota's fascia is lifted out of the abdomen and the posterior dissection is completed under direct vision (**Figure 59.10**). The renal bed is inspected following removal of the kidney. Any remaining lymph nodes on the great vessels are removed and separately labeled for histology, which is essential for staging the tumor. Titanium clips to the renal bed may be needed for further radiotherapy.



59.10

Partial nephrectomy or "nephron-sparing" surgery

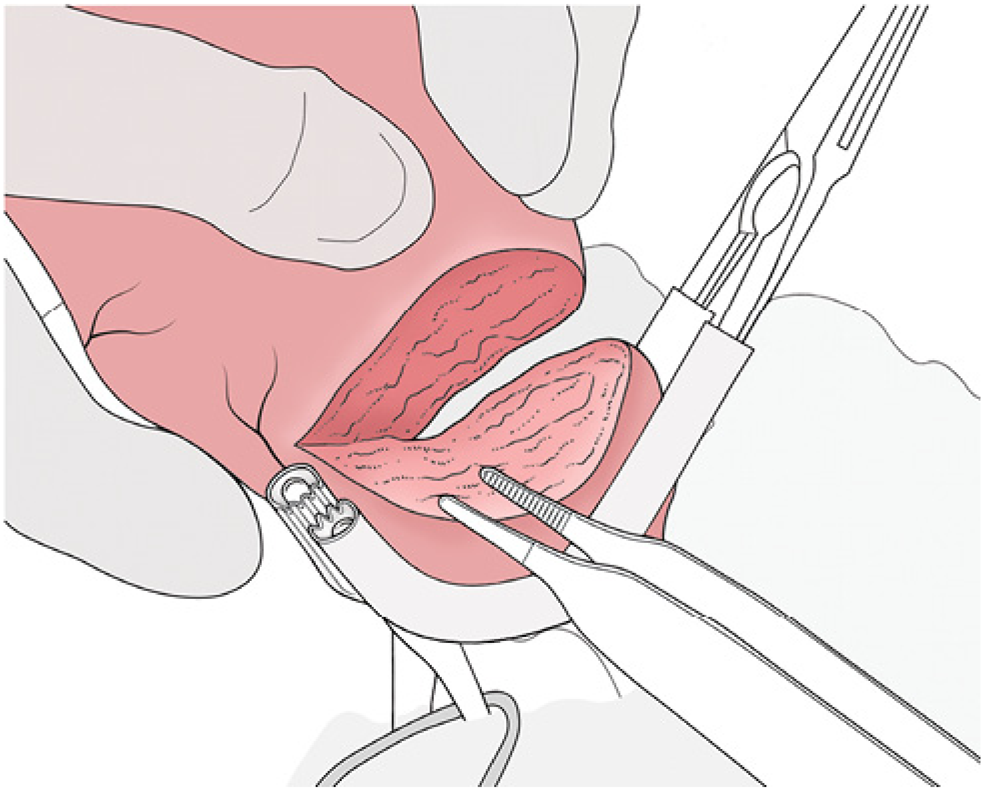
Partial nephrectomy or "nephron-sparing" surgery (NSS) can be achieved in some children. The ideal candidates have single, favorable histology masses, not involving the renal hilum and sparing at least a third of the kidney and no metastatic disease. Surgery is initiated in a similar fashion to conventional nephrectomy. After opening Gerota's fascia and freeing the kidney from the perirenal fat, the tumor can be assessed (**Figure 59.11**). Intraoperative US can be beneficial for determining tumor depth and parenchymal extension and finding unrecognized intrarenal tumors. Biopsies are then taken of perirenal fat and regional lymph nodes for frozen section to establish staging. Finally, the affected kidney is walled off from the peritoneum with laparotomy sponges in case of inadvertent tumor spillage.



59.11

Vascular control of the renal artery should preferably be achieved by compression or clamping of the renal parenchyma because vascular clamping is associated with a higher rate of renal complications. If vascular clamping is adopted, the procedure should be completed within a safe, warm ischemia time window of 30 minutes, which can be extended up to 1 hour by surface cooling with ice slush. A vessel loop is passed around the vascular pedicle to provide rapid access to the blood supply if needed. Circumferential scoring of the planned extent of resection is made

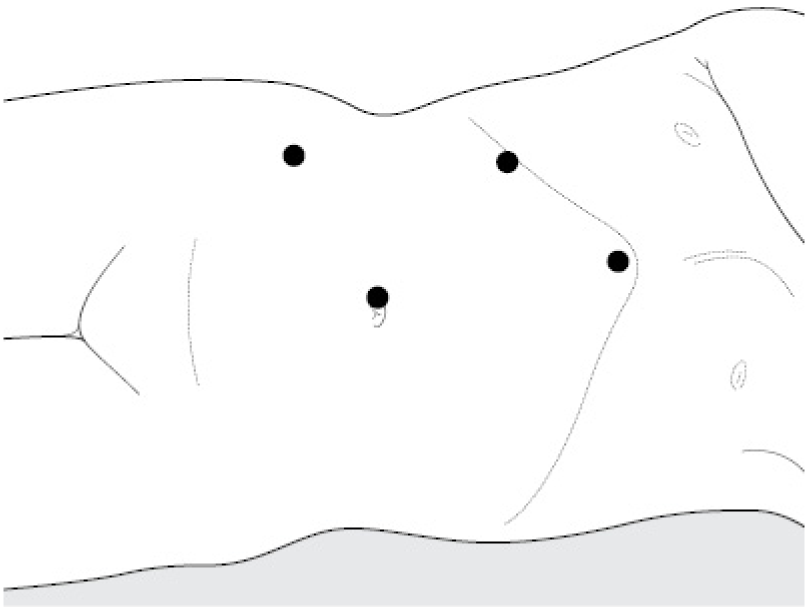
(**Figure 59.12**) and ligation beginning with tumor-feeding vessels is carried out. Frozen sections of the tumor base are obtained, aiming for a resection margin of 0.5–1 cm. A watertight seal of the collecting system is essential to prevent urinary fistula formation. Hemostasis is a crucial element of NSS and various hemostatic agents and sealants can be used in this operation to facilitate hemostasis of the transected renal surface.



59.12

Laparoscopic operation

If the tumor is small and confined to the kidney, laparoscopy offers an alternative to open surgical removal. Three or four ports are typically used (**Figure 59.13**). After gaining access, a stepwise approach follows the same sequences as open surgery including reflecting the colon, entering the retroperitoneal space, mobilization of the kidney, sequential ligation and division of vasculature followed by delivery of the tumor through a Pfannenstiel incision.



59.13

PRE- AND POSTOPERATIVE CHEMOTHERAPY

Unilateral disease

Current regimens include vincristine and actinomycin, administered for 4 weeks, with surgery performed on week 5–6. The type and duration of postoperative chemotherapy is related to the surgical stage and histologic type of tumor. In a completely excised tumor that is confined to the kidney with a low-risk histology, no further treatment may be necessary. Combination treatment with actinomycin and vincristine is usual however, with the option to add doxorubicin with or without abdominal radiotherapy for higher-grade tumors.

Bilateral disease

North American trial data on bilateral diseases advocate for a preoperative three-drug regime (vincristine, dactinomycin, and doxorubicin) and an objective aim to increase NSS to 50%. While the outcomes for bilateral Wilms' have greatly improved, with options for surgical management expanding, overall survival data from North America and Europe still lag behind management of unilateral disease. Decisions on management of bilateral disease are complex and require a team approach.

Metastatic disease

Pulmonary metastases documented on X-ray or CT of the chest preoperatively mandate pre-treatment multi-regimen chemotherapy. This will be followed by further multi-regimen chemotherapy after surgery, with or without radiotherapy, if there has been a poor response.

COMPLICATIONS

The most common intraoperative complication is bleeding. Intraoperative tumor spill is an important complication with a markedly decreased incidence in the SIOP patients, 2.2%, compared to the NWTSG patients, 15% ($P < 0.001$). The most common complication after surgery is small bowel obstruction due to intussusception of the small bowel or adhesions (>5%). Chylous ascites is a less common but significant complication.

CONCLUSION

The management of WT consists of chemotherapy combined with surgery and, less frequently, radiotherapy. Radiotherapy is mainly administered in the presence of high-risk tumors, local spillage, or metastases in the lung that do not respond adequately to chemotherapy. The management of WT includes a team consisting of a surgeon, oncologist, and histopathologist.

FURTHER READING

Cozzi DA, Zani A. Nephron-sparing surgery in children with primary renal tumor: Indications and results. *Semin Pediatr Surg* 2006; 15: 3–9.

Duarte RJ, Dénes FT, Cristofani LM, Srougi M. Laparoscopic nephrectomy for Wilms' tumor. *Expert Rev Anticancer Ther* 2009; 9: 753–61.

Duffy PG, Sebire N. Genitourinary malignancies. In: Thomas D, Duffy PG, Rickwood AMK (eds). *Essentials of Paediatric Urology*. London: Martin Dunitz; 2008: pp. 295–305.

Murphy AJ, Davidoff AM. Bilateral Wilms tumor: A surgical perspective. *Children (Basel)* 2018; 5: 134.

Vujanic GM, Gessler M, Ooms AHAG, et al. The UMBRELLA SIOP-RTSG 2016 Wilms tumour pathology and molecular biology protocol. *Nat Rev Urol* 2018; 15(11): 693–701.

Wilms M. *Die Mischgeschwülste der Niere*. Leipzig: Arthur Georgi; 1899: pp. 1–90.

60

Neuroblastoma

ANDREW M. DAVIDOFF

HISTORY

Neuroblastoma is the most common abdominal malignancy of childhood and the most common cancer in infants. It arises in primitive precursors of the sympathetic nervous system and, consequently, occurs in the adrenal medulla and sites of sympathetic ganglia not only in the abdomen but throughout the body. The annual incidence of neuroblastoma is about one case per 100 000 children, thereby, accounting for about 8% of all malignancies in those younger than 15 years of age. However, neuroblastoma is responsible for approximately 15% of all pediatric cancer deaths. The incidence of neuroblastoma is highest in very young children. Over half of the cases are detected by 18 months of age and approximately three-quarters by 3 years of age; only 5% occur in children over the age of 10 years.

Based on cooperative group studies, neuroblastoma became the first tumor for which clinical and biologic factors predictive of outcome were identified. This allowed not only for more accurate assessment of prognosis but also for the adjustment of therapy intensity, lessening therapy intensity for children with low-risk disease to avoid long-term therapy toxicity and increasing therapy intensity for those with high-risk disease to increase the likelihood of cure. Clinical risk factors include age and stage at diagnosis; biologic risk factors include amplification of the *MYCN* proto-oncogene, as first identified by Seeger in 1985, and tumor histopathology, using an age-linked classification schema first described by Shimada in 1984 (subsequently modified in 2003 in the International Neuroblastoma Pathology

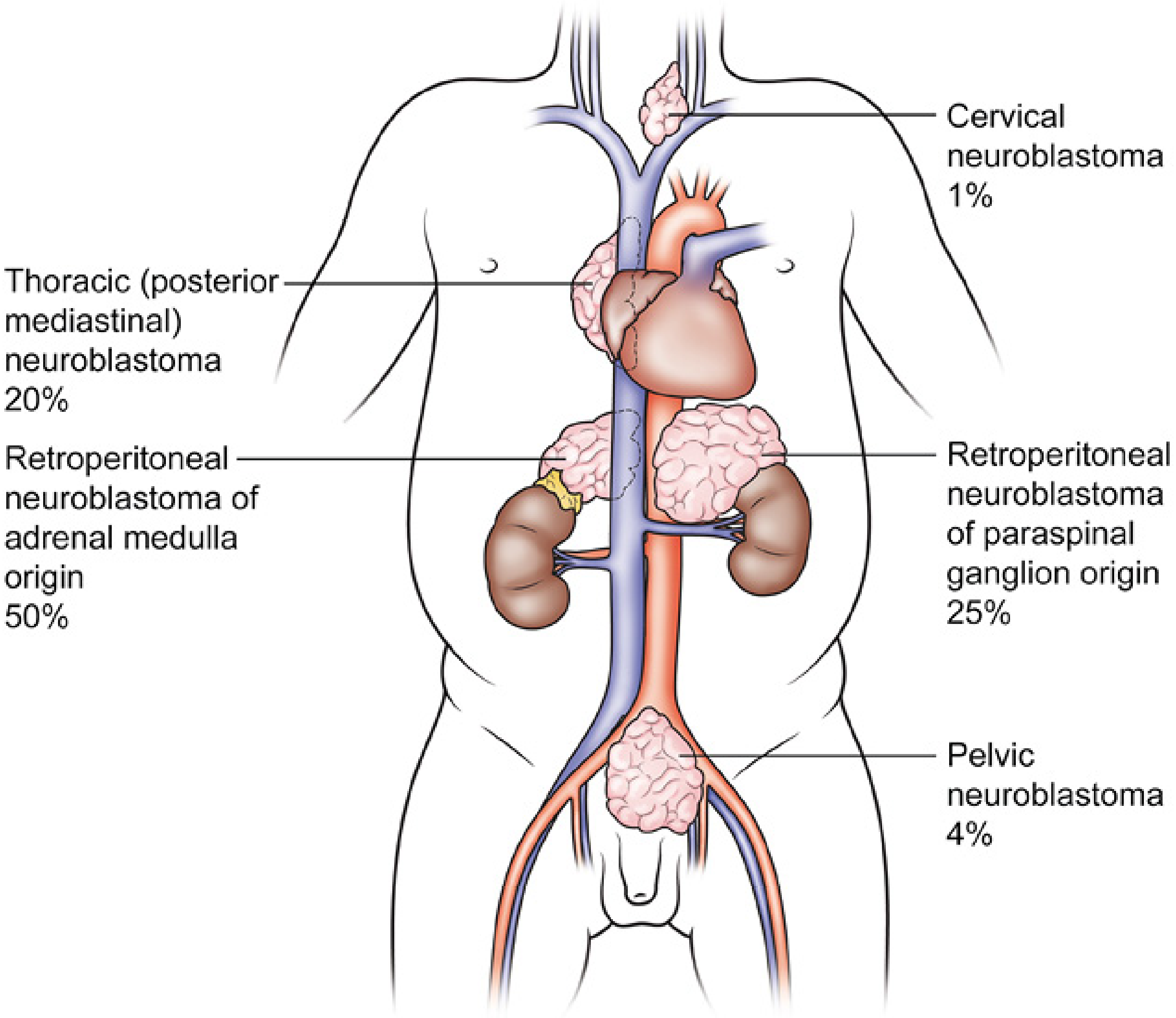
Classification) based on the differentiation grade of the intratumoral neuroblasts, the cellular turnover index (mitosis–karyorrhexis index (MKI)), and the presence or absence of Schwannian stroma. With recent advances in clinical genomics, increasing molecular genetic abnormalities are being identified in tumors (and germline). These are being assessed for independent prognostic significance, contribution to tumor pathogenesis, and suitability for therapeutic targeting.

Complete resection of neuroblastoma offers definitive therapy with a generally excellent outcome for most patients with localized disease, although the benefit of complete tumor removal may be overestimated because of the possibility that localized neuroblastomas may undergo spontaneous maturation or regression. The role of surgery is even less clear in the curative treatment of patients with high-risk neuroblastoma and attempts at resection can be associated with significant complications. However, with the advent of more effective chemotherapy, primary tumors are now rendered smaller, more fibrotic, and more amenable to resection. Consequently, macroscopic clearance of disease is currently a realistic expectation and getting tumors to a state of minimal residual disease may improve the effectiveness of adjuvant therapies such as radiation therapy and immunotherapy.

PRINCIPLES AND JUSTIFICATION

Clinical presentation

Patients with neuroblastoma usually present with signs and symptoms that reflect the primary site and extent of disease, although localized disease can be asymptomatic. As 75% of neuroblastoma occurs in the abdominal cavity, an abdominal mass detected on physical examination is a common clinical feature, as is the complaint of abdominal pain. Other primary sites of neuroblastoma include the posterior mediastinum (20%), the cervical region (1%), and the pelvis (4%) (organ of Zuckerkandel) (**Figure 60.1**). Respiratory distress or dysphagia may reflect a thoracic tumor. Altered defecation or urination may be caused by mechanical compression from a pelvic tumor or by spinal cord compression from a paraspinal tumor. Spinal cord compression may also present as an altered gait. A tumor in the neck or upper thorax can produce Horner syndrome (ptosis, miosis, and anhydrosis), enophthalmos, and heterochromia of the iris. Acute cerebellar ataxia has also been observed, characterized by the dancing-eye syndrome, which includes opsoclonus, myoclonus, and chaotic nystagmus. Two-thirds of these cases occur in infants with mediastinal primary tumors. Additional signs and symptoms that reflect excessive catecholamine or vasoactive intestinal polypeptide (VIP) secretion include hypertension, diarrhea and weight loss.

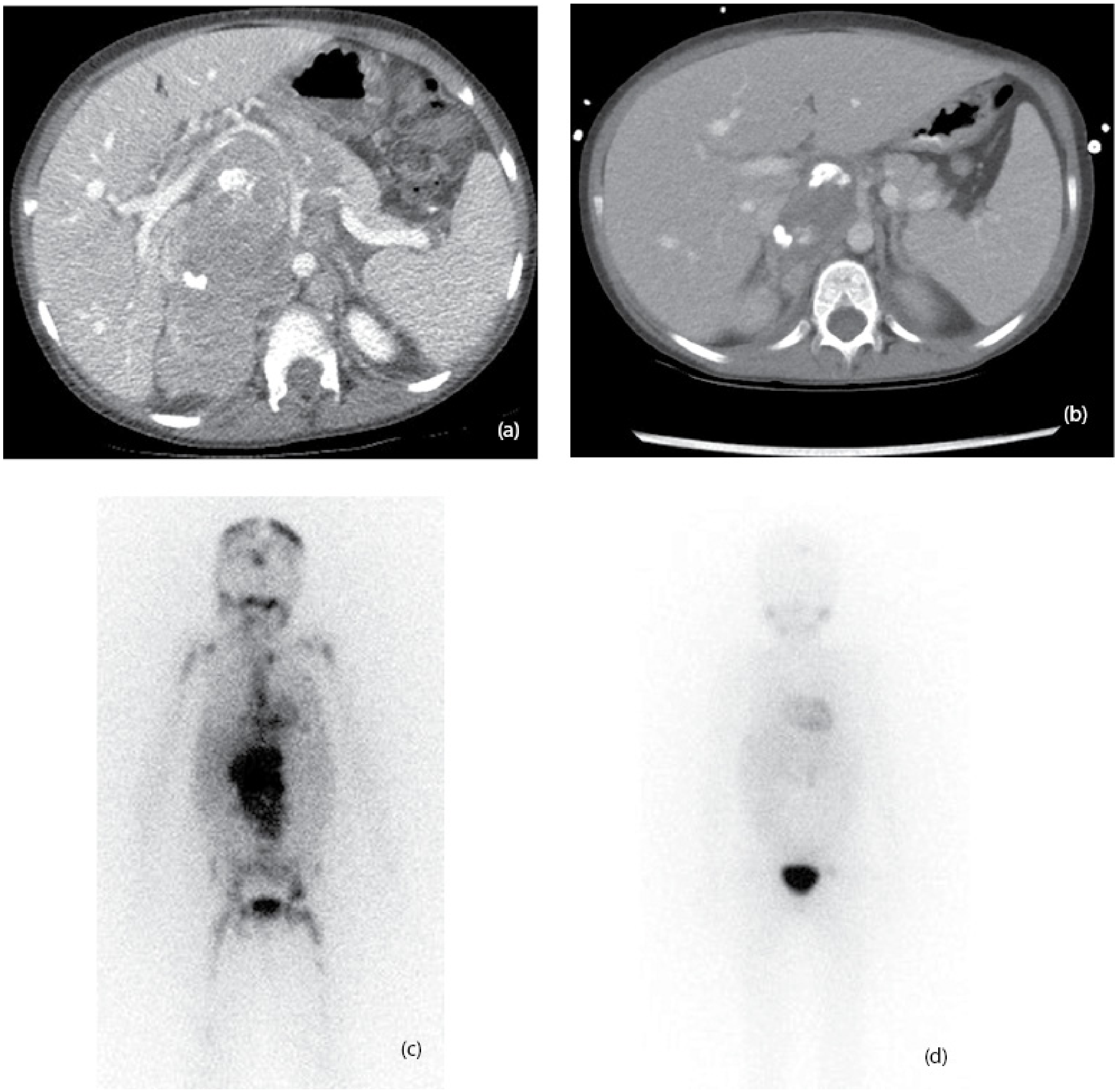

Cervical neuroblastoma 1%
Thoracic (posterior mediastinal) neuroblastoma 20%
Retroperitoneal neuroblastoma of adrenal medulla origin 50%
Retroperitoneal neuroblastoma of paraspinal ganglion origin 25%
Pelvic neuroblastoma 4%

60.1

More than 40% of children with neuroblastoma have metastatic disease at diagnosis. These patients are often quite ill and have systemic symptoms caused by widespread disease. Neuroblastoma has a pattern of metastatic disease to the bone marrow, lymph nodes, and bone. These metastases may manifest as anemia from marrow infiltration, or bone pain and/or limp from cortical metastases. In very young patients, metastatic infiltration of the liver may result in hepatomegaly. Metastatic disease may be also associated with darkened eyes, referred to as "raccoon eyes," resulting from retro-orbital venous plexus spread, or purple, subcutaneous nodules, referred to as "blueberry muffin" lesions.

The diagnosis of neuroblastoma is generally made by histopathologic evaluation of primary or metastatic tumor tissue, or by the demonstration of tumor cells in the bone marrow together with elevated levels of catecholamine metabolites in the urine. Cross-sectional imaging (computed tomography (CT) or magnetic resonance imaging (MRI)) together with ^{131}I-metaiodobenzylguanidine (MIBG) scanning complete the initial imaging workup. The typical appearance of an abdominal neuroblastoma on CT scan or MRI is of an irregular mass arising in the suprarenal or para-aortic region which enhances after the administration of IV contrast medium. Cross-sectional imaging will also help to evaluate the extent of vascular encasement, and serial scanning after therapy will document the response to treatment. MIBG can identify sites of occult metastases. A technetium-99 m methylene diphosphonate (99 mTc-MDP) bone scan can be used when MIBG scanning is unavailable or when a neuroblastoma is not MIBG-avid, as occurs in about 10% of cases. Evaluation of bone marrow aspirates and trephine biopsies are also a critical part of the metastatic workup.

A representative CT scan is shown in **Figure 60.2a** — a 5-year-old boy with stage 4 neuroblastoma. The extent of vascular displacement and envelopment by the tumor is readily apparent. The MIBG scan, shown in **Figure 60.2b**, of the same child, obtained at diagnosis, shows extensive metastatic disease. A scan of the same child after neoadjuvant chemotherapy shows the extent of tumor regression (**Figure 60.2c**) and after surgery, improvement in MIBG scan (**Figure 60.2d**).



60.2a–d

Staging

Staging systems are used to document disease extent and contribute to risk and treatment stratification. The most common such system used until recently has been the International Neuroblastoma Staging System (INSS). It is a surgicopathologic staging system that depends on the completeness of resection of the primary tumor, assessment of ipsilateral and contralateral lymph nodes, and the relation of the primary tumor to the midline. About 33% of patients have stage 1 or 2 disease, 60% have stage 3 or 4 disease, and about 7% present with stage 4S disease.

Although INSS has been shown to have prognostic relevance, there have been some difficulties with its widespread use. The expertise and aggressiveness of the surgeon influence tumor stage, lymph node sampling is done erratically, and patients who are simply observed without surgery cannot be appropriately staged. Therefore, a uniform, pre-treatment staging system that could be used easily throughout the world, and subject to real-time central review, was developed (International Neuroblastoma Risk Group Staging System (INRGSS)) based on tumor imaging rather than the extent of surgical resection. In this staging system, detailed in **Table 60.1**, localized tumors are staged based on the absence (L1) or presence (L2) of one or more of 20 image-defined risk factors (IDRFs, see **Table 60.2**) which generally reflect encasement of vital structures, primarily vessels and nerves, as determined by diagnostic imaging studies, and which are associated with a lower complete resection rate and a greater risk of surgery-related complications.

Table 60.1 **International Neuroblastoma Risk Group staging system**

Stage	Description
L1	Localized tumor not involving vital structures (as defined by image-defined risk factors) and confined to one body compartment
L2	Locoregional tumor with presence of one or more image-defined risk factors
M	Distant metastatic disease (except stage MS)
MS	Metastatic disease in children younger than 18 months with metastases confined to skin, liver, and/or bone marrow

Table 60.2 **Image-defined risk factors for primary resection of localized neuroblastoma**

Anatomic region	Description
Multiple body compartments	Ipsilateral tumor extension within two body compartments (i.e. neck and chest, chest and abdomen, or abdomen and pelvis)
Neck	Tumor encasing carotid artery, vertebral artery, and/or internal jugular vein
	Tumor extending to skull base
	Tumor compressing trachea
Cervicothoracic junction	Tumor encasing brachial plexus roots
	Tumor encasing subclavian vessels, vertebral artery, and/or carotid artery
	Tumor compressing trachea
Thorax	Tumor encasing aorta and/or major branches
	Tumor compressing trachea and/or principal bronchi
	Lower mediastinal tumor infiltrating costovertebral junction between T9 and T12 (may involve the artery of Adamkiewicz supplying the lower spinal cord)
Thoracoabdominal junction	Tumor encasing aorta and/or vena cava
Abdomen and pelvis	Tumor infiltrating porta hepatis and/or hepatoduodenal ligament
	Tumor encasing branches of superior mesenteric artery at mesenteric root
	Tumor encasing origin of celiac axis and/or origin of superior mesenteric artery
	Tumor invading one or both renal pedicles
	Tumor encasing aorta and/or vena cava
	Tumor encasing iliac vessels
	Pelvic tumor crossing sciatic notch
Intraspinal tumor extension	Intraspinal tumor extension (whatever the location) provided that more than one-third of spinal canal in axial plane is invaded, the perimedullary leptomeningeal spaces are not visible, or the spinal cord signal intensity is abnormal
Infiltration of adjacent organs and structures	Pericardium, diaphragm, kidney, liver, duodenopancreatic block, and mesentery

Treatment

Increasing evidence indicates that the biologic and molecular features of neuroblastoma are highly predictive of clinical behavior. Therefore, neuroblastoma has served as a paradigm for phenotypic risk assessment and treatment assignment. The predictive value of these biologic factors is important not only for the oncologist when considering appropriate chemotherapy, but also for the surgeon when considering the timing and extent of an operative resection in a child with neuroblastoma. The variables used for risk stratification in recent Children's Oncology Group (COG) clinical trials served to categorize infants and children with neuroblastoma in three risk groups predictive of relapse: Low, intermediate, and high risk. The probability of prolonged disease-free survival for patients in each group was >95%, >90%, and <30%, respectively.

More recently, the INRG classification system is increasingly being used in which survival tree regression analyses with event-free survival (EFS) as the primary end point determined that seven prognostic variables could define 16 different pre-treatment risk groups (**Table 60.3**). These risk groups generate four categories based on expected 5-year EFS: Very low (>85% EFS, 28.2% of patients), low (>75 to ≤85% EFS, 26.8% of patients), intermediate (≥50 to 75% EFS, 9.0% of patients), and high (<50% EFS, 36.1% of patients) risk.

Table 60.3 International Neuroblastoma Risk Group (INRG) pre-treatment classification

INRG stage	Age (months)	Histologic category	Grade of tumor differentiation	MYCN	11q aberration	Ploidy	Pre-treatment risk group
L1/L2		GN maturing; GNB intermixed					A Very low
L1		Any, except GN maturing or GNB intermixed		NA			B Very low
				Amp			K High
L2	<18	Any, except GN maturing or GNB intermixed		NA	No		D Low
					Yes		G Intermediate
	>18	GNB nodular; neuroblastoma	Differentiating	NA	No		E Low
					Yes		H Intermediate
			Poorly differentiated or undifferentiated	NA			
				Amp			N High
M	<18			NA		Hyperdiploid	F Low
	<12			NA		Diploid	I Intermediate
	12 to <18			NA		Diploid	J Intermediate
	<18			Amp			O High
	>18						P High
MS	<18			NA	No		C Very low
					Yes		Q High
							R High

Neither adjuvant chemotherapy nor radiation therapy appears to be necessary for the management of most patients with very low- and low-risk disease; surgical resection is generally the only therapy given to these patients. Increasingly, as risk factors are further refined, patients with very low- or low-risk disease are simply being observed without resection of the primary tumor.

The overall surgical goal in intermediate-risk patients is to perform the most complete tumor resection possible, consistent with preservation of full organ and neurologic function. This may necessitate leaving residual disease adherent to critical structures. If a primary tumor is judged by the surgeon to be unresectable, a diagnostic biopsy is generally obtained and chemotherapy initiated with surgery being delayed.

The general approach to treating patients with high-risk neuroblastoma includes intensive induction chemotherapy, myeloablative consolidation therapy with stem cell rescue, and targeted therapy, including differentiating agents and immunotherapy, for minimal residual disease. Resection of the primary tumor and bulky locoregional metastatic sites is generally attempted after the fifth cycle, although it may be performed earlier, given that further tumor shrinkage is generally minimal after the second or third cycle.

OPERATION

Early surgery is appropriate for localized tumors where resection can be undertaken safely, unless the patient qualifies for an observation-only approach. For those with locally advanced or disseminated disease, the initial use of neoadjuvant chemotherapy followed by surgery allows for a more complete resection.

The operation, as described by Edward Kiely in a previous edition of this textbook and in manuscripts listed at the end of this chapter, was developed to deal with the problem of vascular encasement. It also allows a planned and systematic approach to tumor resection and minimizes the risk of vascular accidents while achieving complete tumor clearance.

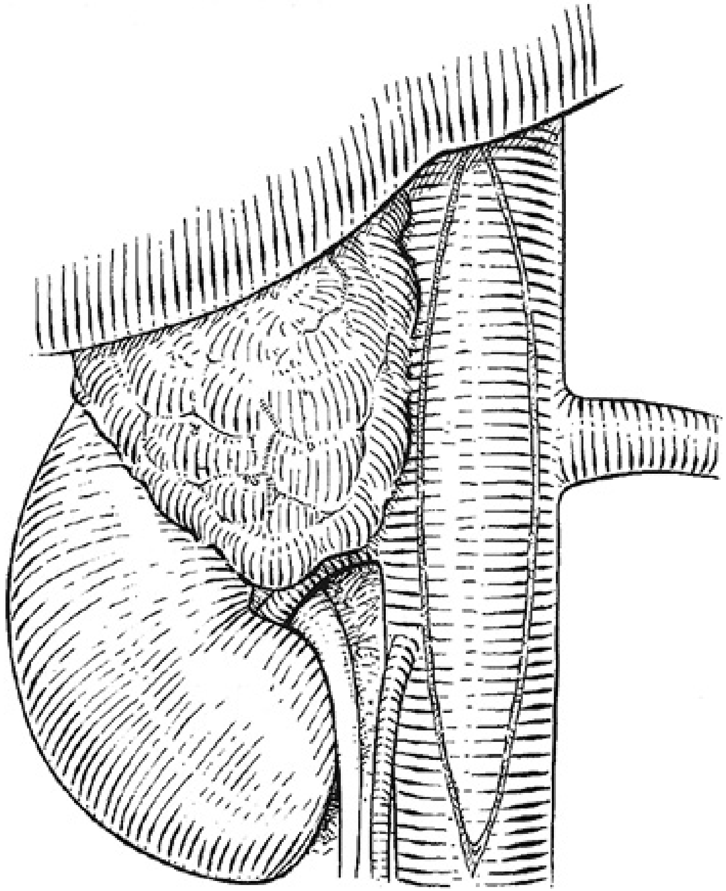
As neuroblastoma rarely invades the tunica media of major blood vessels, the dissection may be performed in the subadventitial plane with a scalpel. The success of the procedure depends entirely on the attitude and persistence of the surgeon. The operation consists of three phases: Vessel display, vessel clearance, and tumor removal. Vessel display is the effort to expose the circumference of each blood vessel that traverses the tumor, in continuity. If this can be accomplished, these vessels can generally be cleared of tumor. Once the vessels are free, the tumor can be removed piecemeal. The order in which the vessels are displayed and cleared depends on the site and size of the tumor together with the surgeon's preference.

Open operation

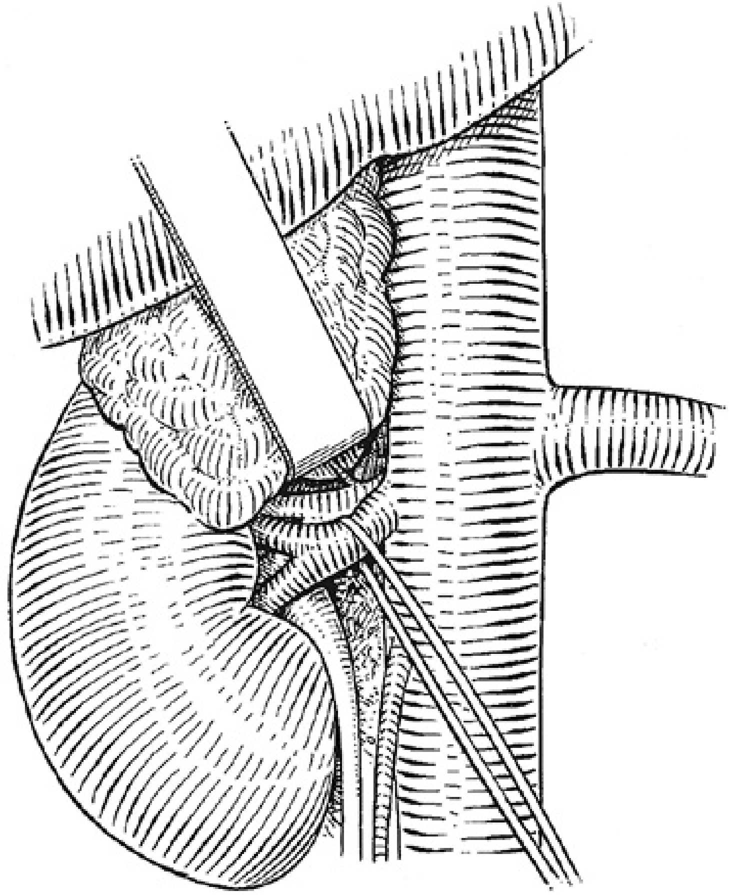
Briefly, there are five major points to safe resection of neuroblastomas associated with one or more image-defined risk factor.

- Approach the operation as a vascular-type operation in which identification and skeletonization of the major intra-abdominal vessels is critical.
- The tumor should be removed piecemeal. In particular, undue torque on the renal artery in an effort to clear tumor from behind the renal hilum in one piece may result in injury to the intima of the artery, with resultant vessel spasm and/or thrombosis, leading to renal ischemia.
- Dissection commences distal to the lower edge of the tumor, generally along the common or external iliac artery, and proceeds proximally to encounter the tumor along the aorta, identifying the major arterial branches (and left renal vein).
- Tumors involving the structures of the porta hepatis and the pelvis can be particularly difficult.
- Thoracic tumors are usually more easily cleared from the vascular structures, but they often extend into the intervertebral foramina. Extraction of tumor from this location is of uncertain benefit and can be associated with significant complications.

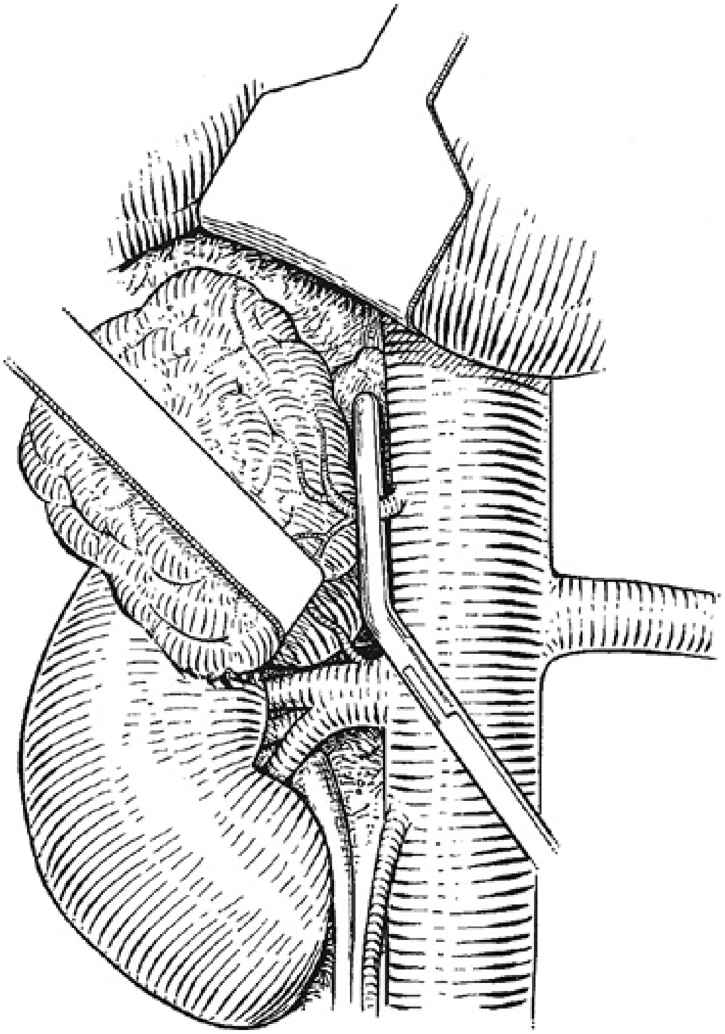
For a localized, L1 tumor (**Figure 60.3**), the abdomen is opened through a transverse supraumbilical incision. For right-sided tumors, the ascending colon and duodenum are reflected medially. The inferior vena cava below the limit of the tumor is exposed by incision of the tunica adventitia longitudinally along the middle of the vessel. The dissection advances proximally, exposing the anterior wall in the 12 o'clock position. If tumor is encountered anterior to the vena cava, it is incised down to the tunica media. This dissection is advanced proximally until the liver is reached. Subsequently, the right renal vein is displayed in similar fashion (**Figure 60.4**). The cava is then cleared of tumor and elevated to expose the right renal artery. Once the right renal artery and vein are displayed, the cava above the renal vein may be cleared of tumor until the right adrenal vein is reached (**Figure 60.5**). For left-sided tumors the approach is similar but with mobilization of the descending colon. Often, exposure is best with medial mobilization of the spleen and pancreas. The medial dissection is along the left lateral edge of the aorta.



60.3



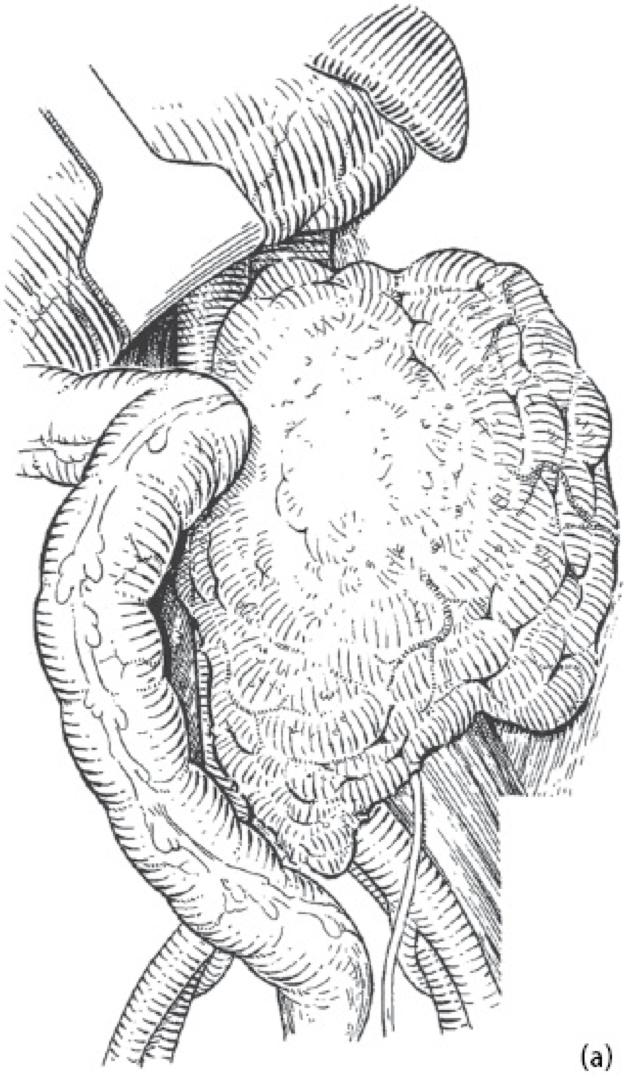
60.4



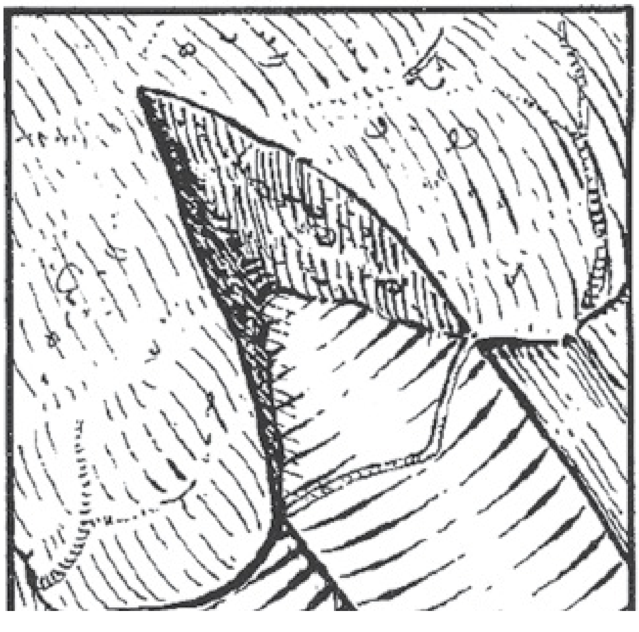
60.5

More commonly, the surgeon is faced with an L2 lesion with extensive residual tumor remaining following intensive chemotherapy. A key feature of the operation involves the systematic dissection of major vessels which abut or traverse the tumor. Dissection is commenced below the tumor where the vessel emerges — usually an iliac vein on the right or iliac artery on the left side. All tissue is divided under tension produced by the assistant and the surgeon picking up tunica adventitia and retracting laterally. The knife then divides the tissue down to the tunica media in a perpendicular direction. The dissection commences where the vessel is normal and not surrounded by tumor. The dissection then continues proximally, dividing tumor and adventitia and keeping the vessel wall in view all the time. The technique remains the same — division of tissue under tension in lengths of 1–2 cm at a time. The dissection moves from what is visible to what is not visible. Hemostasis is achieved using bipolar diathermy. In general, the tissue is divided first. Initial coagulation may obliterate tissue planes and make the procedure more difficult.

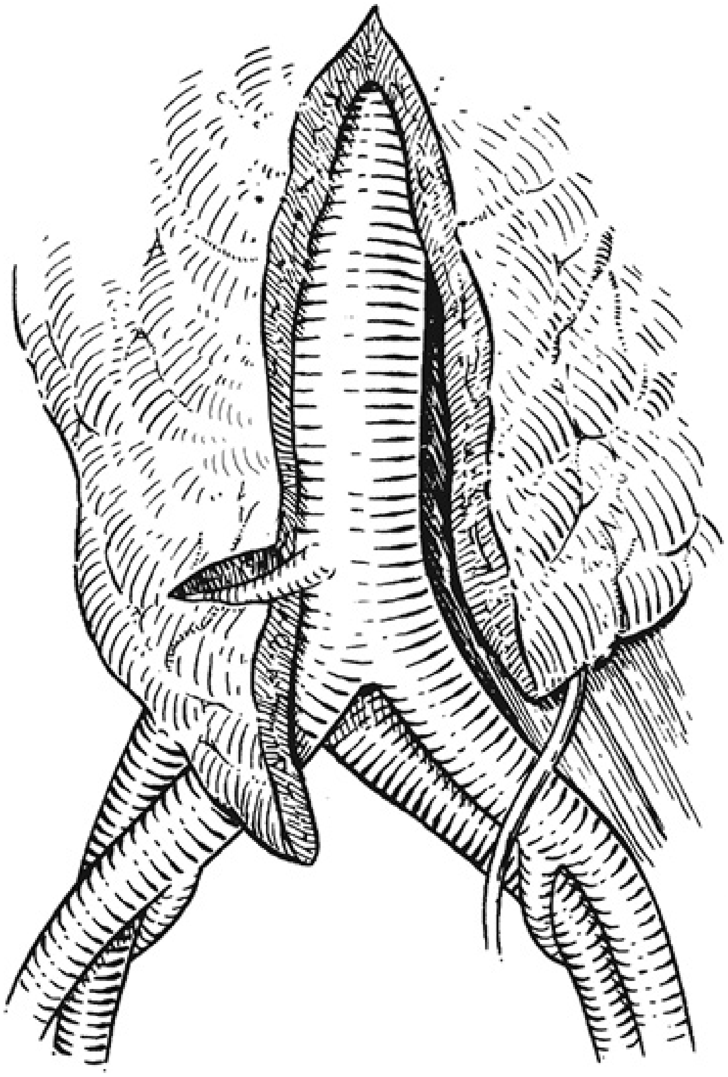
For extensive retroperitoneal and left-sided tumors, the descending and sigmoid colon are mobilized, together with the spleen, pancreas, and stomach, and placed in an intestinal bag (**Figure 60.6a**). The dissection commences just distal to the tumor edge along the middle of whichever artery is present (**Figure 60.6b**). The dissection frequently commences at the left common iliac artery, exposing the subadventitial layer. The dissection then advances proximally, dividing the tumor and adventitia down to the tunica media of the vessel (**Figure 60.7**). The dissection continues proximally until the aorta is reached and then progresses upward along this vessel. The inferior mesenteric artery is soon encountered and the division of the tumor and adventitia continued until the left renal vein is reached. Once the renal vein is encountered, a segment of 4–5 cm of the vein is cleared so that it can be retracted. Cautious dissection will bring the origin of the left renal vein into view. The plane of dissection then changes from the 12 o’clock position to the 2 o’clock position to avoid the superior mesenteric and celiac arteries. The operation proceeds until the superior limit of the tumor is reached and this may entail division of the median arcuate ligament to enter the posterior mediastinum.



(a)

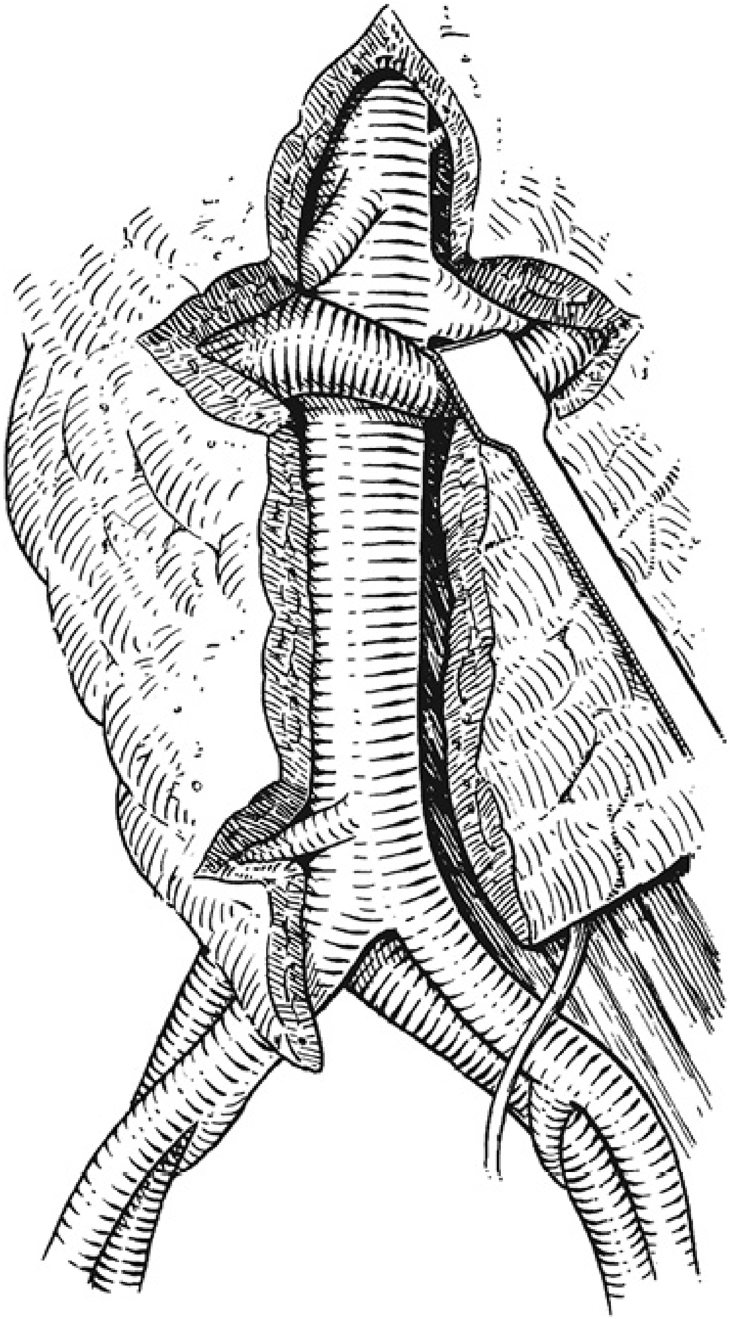


60.6a,b

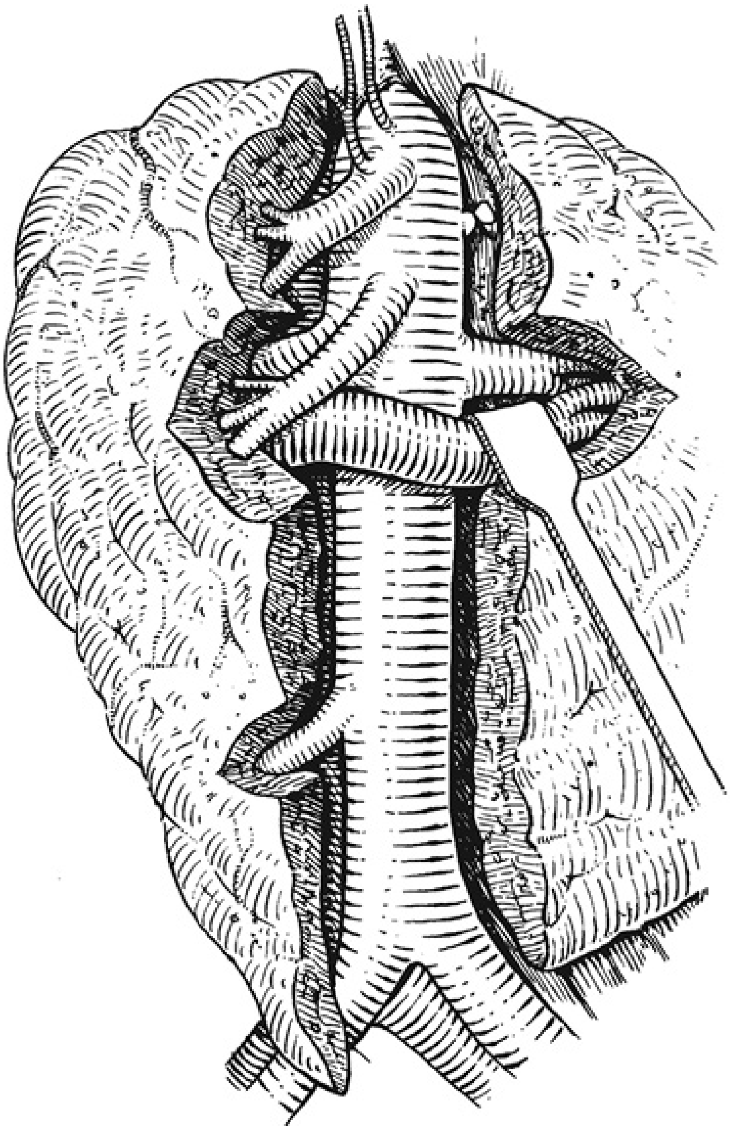


60.7

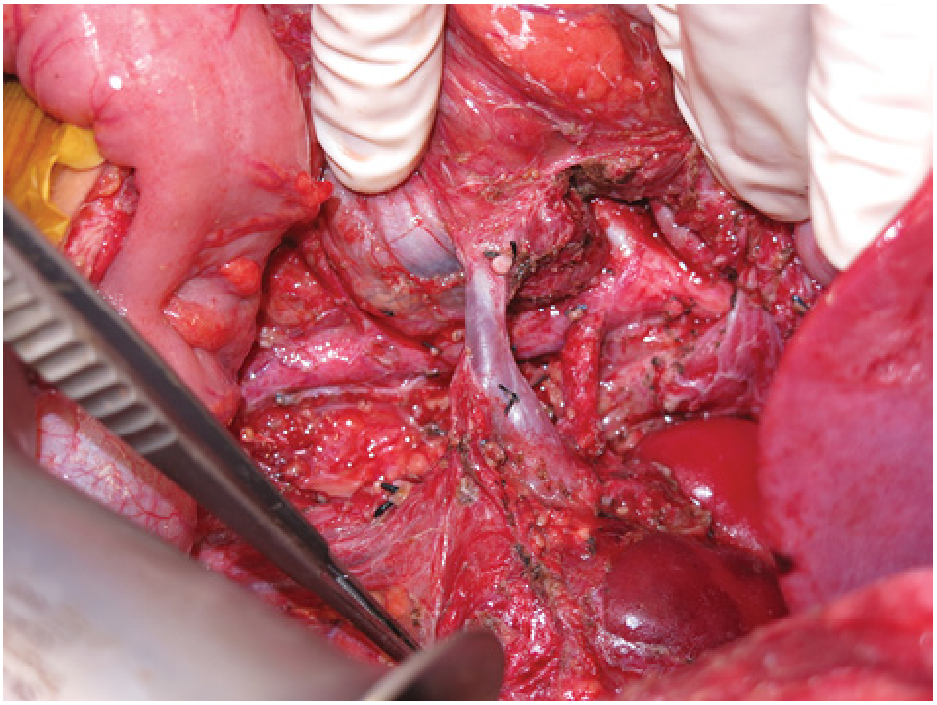
Each of the encased visceral arteries is dissected in turn — left renal, superior mesenteric, and celiac (**Figure 60.8**). The dissection advances distally on these vessels as far as is necessary. It is not unusual to have to dissect left gastric, hepatic, and splenic arteries for some distance. Once these vessels are safe and in view, they may be cleared circumferentially of tumor prior to removal of the residual tumor (**Figure 60.9**). The intraoperative photograph in **Figure 60.10** shows the final appearance of the operative field showing the left renal vein, and aorta and associated branches (inferior mesenteric artery, left renal artery, superior mesenteric artery, celiac axis) completely cleared of tumor.



60.8



60.9



60.10

Minimally invasive surgery

Increasingly, minimally invasive approaches (laparoscopic or thoracoscopic) are being used for neuroblastoma resections. Because piecemeal removal of neuroblastoma and resections that leave microscopic, and in some cases macroscopic, residual disease do not appear to influence patient outcome or violate oncologic principles, the decision to use a minimally invasive approach to resection is often dictated by the comfort and experience of the surgeon, rather than image-defined risk factors. Most L1 lesions, particularly in the chest, should be amenable to a minimally invasive resection and even L2 can be considered for a minimally invasive approach, depending on the skill of the surgeon. Generally, the principles of the conduct of the operation — mobilization of the mesentery, exposure, vascular control — should be followed whether using an open or a minimally invasive approach.

POSTOPERATIVE CARE

Postoperative care follows the same principles as for any other major laparotomy. Intravascular monitoring is continued for 24–48 hours. Urine output is carefully measured, and intravascular volume adjusted accordingly, as significantly third-space loss is common during a long resection of a complex abdominal neuroblastoma. Effective pain relief is delivered in the form of either epidural infusion or IV opiate infusion. Enteral feeding usually resumes after about 3 days. Diarrhea is not uncommon when celiac and superior mesenteric arteries have been cleared of tumor.

COMPLICATIONS

Due to the extensive surgery often required, intraoperative and postoperative complications are not uncommon. As many as 80% of patients will experience significant blood loss that requires transfusion either in the operating room or in the early postoperative period. Up to 10% will suffer an injury to a major vascular structure (aorta, vena cava, or renal vessels). Injury to other viscera (stomach, bowel, liver, spleen, or kidney) occurs in approximately 5% of cases. On occasion, this necessitates removal of the injured organ, with the kidney being the most common. Postoperative complications are variable. Wound complications occur in 1–5% of cases, as does postoperative bowel obstruction. In addition, hypertension, chyle leak into the thorax or abdomen, pleural effusion, infection and sepsis, diarrhea, and the need for prolonged total parenteral nutrition have been known to occur. Rarely, a patient will have to be emergently re-explored for postoperative hemorrhage or bowel obstruction.

OUTCOME

The surgical morbidity after this operation is similar to that encountered after other major procedures. Wound dehiscence or incisional hernias occur in less than 1% of cases.

Chylous ascites is not unusual in the days following operation, but it generally resolves without the need for specific treatment. On rare occasions, bowel rest with parenteral nutrition may be required.

Operative mortality is 1%. The effects of age, tumor stage, and changes in chemotherapy protocols confuse attempts to analyze the influence of surgery on the long-term prognosis.

For many years, it has not been clear whether surgical excision contributes to survival in those with metastatic disease. However, as there has been no prospective evaluation of the role of surgery, it cannot be assumed that those who undergo successful surgery necessarily have the same type of disease as those in whom resection was unsuccessful. More recent results suggest a trend toward improved survival where surgery has been complete or near-complete.

FURTHER READING

Adkins ES, Sawin R, Gerbing RB, et al. Efficacy of complete resection for high-risk neuroblastoma: A Children's Cancer Group study. *J Pediatr Surg* 2004; 39(6): 931–6.

Baker DL, Schmidt ML, Cohn SL, et al. Outcome after reduced chemotherapy for intermediate-risk neuroblastoma. *N Engl J Med* 2010; 363(14): 1313–23.

Cecchetto G, Mosseri V, De Bernardi B, et al. Surgical risk factors in primary surgery for localized neuroblastoma: The LNESG1 study of the European International Society of Pediatric Oncology Neuroblastoma Group. *J Clin Oncol* 2005; 23(33): 8483–9.

Cohn SL, Pearson AD, London WB, et al. The International Neuroblastoma Risk Group (INRG) classification system: An INRG Task Force report. *J Clin Oncol* 2009; 27(2): 289–97.

Kiely EM. The surgical challenge of neuroblastoma. *J Ped Surg* 1994; 29: 128–33.

Kiely EM. A technique for excision of abdominal and pelvic neuroblastomas. *Ann R Coll Surg Engl* 2007; 89(4): 342–8.

Monclair T, Brodeur GM, Ambros PF, et al. The International Neuroblastoma Risk Group (INRG) staging system: An INRG Task Force report. *J Clin Oncol* 2009; 27(2): 298–303.

Nuchtern JG, London WB, Barnewolt CE, et al. A prospective study of expectant observation as primary therapy for neuroblastoma in young infants: A Children's Oncology Group study. *Ann Surg* 2012; 256(4): 573–80.

Strother DR, London WB, Schmidt ML, et al. Outcome after surgery alone or with restricted use of chemotherapy for patients with low-risk neuroblastoma: Results of Children's Oncology Group study P9641. *J Clin Oncol* 2012; 30(15): 1842–8.

von Allmen D, Davidoff AM, London WB, et al. Impact of extent of resection on local control and survival in patients from the COG A3973 study with high-risk neuroblastoma. *J Clin Oncol* 2017; 35(2): 208–16.

61

Rhabdomyosarcoma

GIDEON SANDLER AND ANDREA HAYES-JORDAN

HISTORY

Rhabdomyosarcoma (RMS) is the third most common extracranial solid organ tumor of childhood after neuroblastoma and Wilms' tumor and a member of the malignant small round blue cell tumor group. It is thought to arise from embryonic mesenchyme and can occur anywhere in the body. Prognosis is determined by age, site of origin, tumor size and resectability, histopathologic subtype, molecular characteristics, and presence of regional and distant metastases. Clinical outcomes for high-risk tumors remain poor but for low- and intermediate-risk tumors have improved dramatically. Effective neoadjuvant chemotherapy and radiotherapy (RT) have decreased the extent and morbidity of once radical and mutilating operations. Advances in molecular biology are aiding in the development of new effective chemotherapeutic agents.

PRINCIPLES AND JUSTIFICATION

Rhabdomyosarcoma (RMS) accounts for 2–4.5% of all childhood cancer with an incidence of 4.5 cases per million children. It has a bimodal age distribution, reflecting its two main histological subtypes. Embryonal rhabdomyosarcoma (ERMS) accounts for 57% cases and is more common in younger children and males. Older children are affected equally by ERMS and alveolar rhabdomyosarcoma (ARMS), though with a lower incidence overall, the latter accounting for 23% cases. Other rarer types include pleomorphic (anaplastic) RMS and spindle-cell RMS. Most cases are sporadic. Genetic disorders that predispose to RMS include Li Fraumeni syndrome (*p53* mutation), neurofibromatosis type I (*NF1* mutation), *DICER I* mutations, Costello syndrome (germline *HRAS* mutation), Beckwith–Wiedemann syndrome (*11p15.5* mutation), Gorlin syndrome (*PTCH1* mutation) and Noonan syndrome. Other pathogenic genes include loss of *11p15* heterozygosity, gains in chromosome 8, *NRAS* mutations, and *NMYC* and *CDK4* amplification. Approximately 75% of ARMS is associated with *PAX3–FOXO1* or *PAX7–FOXO1* gene fusions.

RISK STRATIFICATION

Risk stratification of a newly diagnosed patient guides therapy and predicts outcome. RMS is stratified into high-, intermediate- and low-risk groups based on the histological subtype, tumor site, pre-treatment stage, and the post-resection/biopsy "clinical group" (**Table 61.1**).

Table 61.1 Rhabdomyosarcoma risk stratification

Risk group	Pre-treatment stage[a]	Clinical group	Site[b]	Histology
Low 1	1 or 2 1	I or II III	Favorable or unfavorable Orbit	Embryonal
Low 2	1 3	III I or II	Favorable Unfavorable	Embryonal
Intermediate	2 or 3 1–3	III I–III	Unfavorable Favorable or unfavorable	Embryonal Alveolar
High	4 4	IV IV	Favorable or unfavorable Favorable or unfavorable	Embryonal Alveolar

[a] Pre-treatment stage dependent on site of disease.

[b] Favorable sites: Genitourinary, biliary tract, non-parameningeal head and neck, orbit.

Age

Though age is not strictly part of risk stratification, children aged 1–9 years (5-year failure-free survival (FFS) 81%) have a better prognosis than those less than 1 year old (5-year FFS 57%) or over 10 years of age (5-year FFS 68%). This is because children less than 1 year old tolerate chemotherapy and RT poorly and are frequently under-dosed, and older children have body surface area-based dose limits on chemotherapy and tend to have unfavorable tumor characteristics such as alveolar histology, nodal positivity, and distant metastases.

Histological diagnosis

ERMS tends to have a more favorable prognosis. Most cases occur in children under 5 years old. The head and neck and genitourinary tract account for about half of the cases each. An even more favorable subtype is the botryoidal variant, which appears grape-like and originates from mucosal-lined hollow viscera and is typical for vaginal, nasopharyngeal, and biliary RMS.

ARMS occurs more frequently in older children and adolescents with a mean age of presentation between 6 and 9 years. It typically appears in the trunk and extremities. Its diagnosis confers a poorer prognosis and by definition the patient cannot be low risk.

Pre-treatment stage

Staging is a preoperative clinical and radiological evaluation based on the TNM system. It is determined by the size (> or <5 cm) and site (favorable versus unfavorable) of the tumor, degree of anatomic extension (T), determination of nodal status (N), and presence or absence of distant metastases (M) (**Table 61.2**).

Table 61.2 **Tumor node metastasis (TNM) pre-treatment staging classification**

Stage	Sites	T	Size	N	M
1^a	Orbit	T_1 or T_2	a or b	N_0 or N_1 or N_x	M_0
	Head and neck (excluding parameningeal)				
	Genitourinary (non-bladder/non-prostate)				
2^b	Bladder/prostate	T_1 or T_2	a	N_0 or N_x	M_0
	Extremity				
	Cranial				
	Parameningeal				
	Other (includes trunk, retroperitoneum, etc.)				
3^b	Bladder/prostate	T_1 or T_2	a	N_1	M_0
	Extremity		b	N_0 or N_1 or N_x	
	Cranial				
	Parameningeal				
	Other (includes trunk, retroperitoneum, etc.)				
4	Any site	T_1 or T_2	a or b	N_0 or N_1	M_1

Definitions

Tumor T $(\text{site})_1$, confined to anatomic site of origin (non-invasive); (a) ≤5 cm in diameter, (b) >5 cm in diameter

T $(\text{site})_2$, extends beyond the organ/tissue of origin and/or fixed to surrounding tissue; (a) ≤5 cm in diameter, (b) >5 cm in diameter

Regional nodes N_0: Regional lymph nodes not clinically involved

N_1: Regional lymph nodes clinically involved

N_x: Clinical status of nodes unknown

Metastasis M_0: No distant metastases

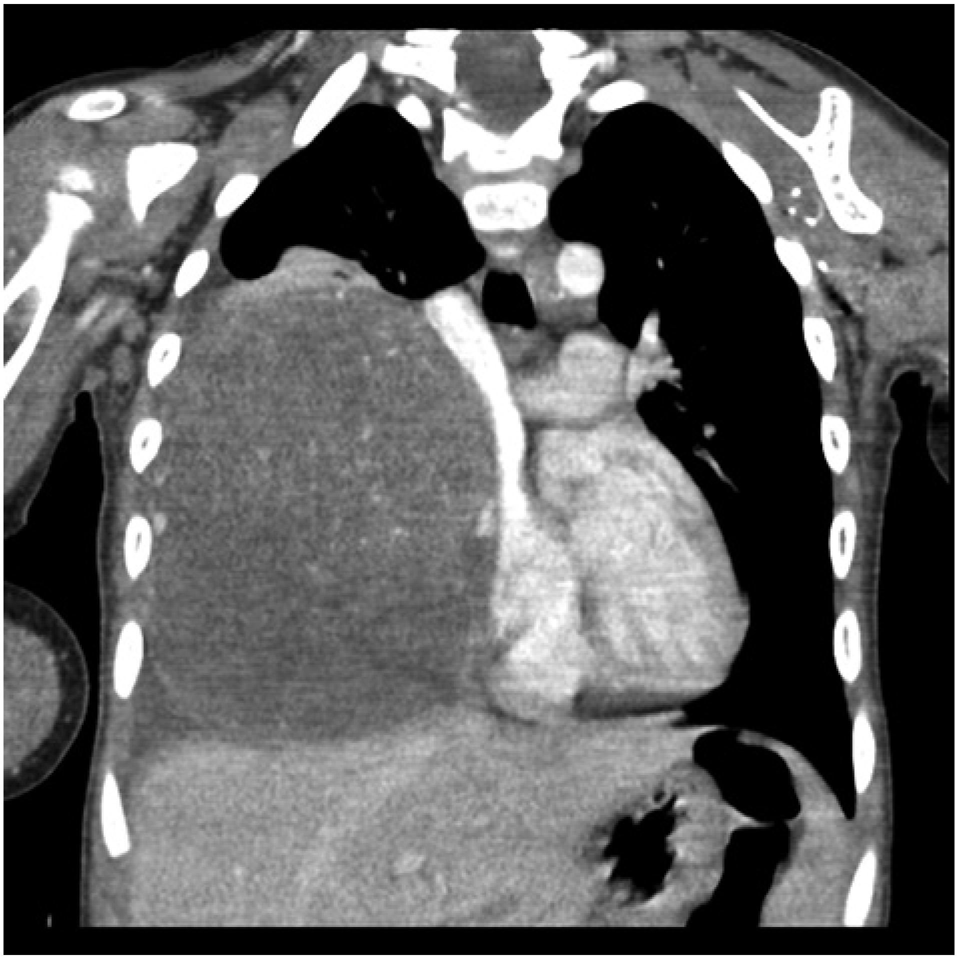
M_1: Metastases present

[a] Favorable site.

[b] Unfavorable site.

Evaluation of local disease usually includes magnetic resonance imaging (MRI), which gives good definition of the tumor itself (particularly in the limbs and trunk) and its relationship with surrounding structures, and computed tomography (CT), which better defines bony involvement, regional nodal status, and the presence of distant metastases (**Figure 61.1**).

Bone scan, bone marrow aspirate and lumbar puncture are also required. ^{18}F-fluorodeoxyglucose positron emission tomography (PET) may offer better definition of regional nodal and occult distant metastases as well as residual tumor post-resection but does not confer prognostic advantages over anatomical imaging and it is not a formal part of the evaluation of RMS patients.



61.1 CT scan of the chest in coronal section demonstrating a rhabdomysarcoma of the right chest wall with intrathoracic extension causing flattening of the right hemidiaphragm, mediastinal shift, and displacement of the right lung in a 3-year-old patient.

Size

Generally, children with smaller tumors (<5 cm) have a more favorable prognosis than those with larger tumors (>5 cm) because of their resectability and lack of metastases. A proportional comparison with body surface area may be a more appropriate prognostic indicator in infants, though this has not been confirmed.

Site

Favorable sites include the genitourinary tract (including paratesticular but excluding bladder, prostate, and kidney), biliary tract, orbit, and non-parameningeal head and neck regions. Unfavorable sites are any site other than those that are favorable and include parameningeal head and neck sites, the extremities, trunk, abdomen, pelvis, bladder/prostate, and retroperitoneum.

Clinical group

A clinical group is assigned based on the completeness of surgical excision, the determination of nodal status and the presence or absence of distant metastases following pathological examination of the operative specimens (**Table 61.3**). Five-year survival for Groups I, III, and III is approximately 90%, 80%, and 70% respectively.

Table 61.3 Rhabdomyosarcoma clinical group

Clinical group	Description
I	Localized disease; tumor removed with a circumferential margin of normal tissue; no nodal involvement
II	Total gross resection with residual microscopic disease at the primary site or draining nodal basin (i.e. positive lymph nodes)
III	Biopsy only or incomplete resection with gross residual disease
IV	Distant metastatic disease

BIOLOGY AND PATHOLOGY

Embryonal rhabdomyosarcoma

ERMS has phenotypical and biological features of embryonic skeletal muscle with a variable number of rhabdomyoblasts and primitive mesenchymal cells in various stages of myogenesis. Botryoidal ERMS is characterized by linear aggregates of tumor cells that tightly abut an epithelial surface. Immunohistochemical markers of ERMS reflect its degree of skeletal muscle differentiation and include vimentin, desmin, actin, creatine kinase M, antibodies against MyoD, and myogenin (weakly).

Alveolar rhabdomyosarcoma

ARMS is characterized by a monomorphous cellular population separated by fibrovascular septae giving the appearance of pulmonary alveoli. Histopathological staining is similar to ERMS except for myogenin, which is usually diffuse and strong. Cytogenetic studies demonstrate typical translocations including t(2;13)(*q35*;*q14*) and t(1;13)(*p36*;*q14*) corresponding to fusion between *FOXO1* and *PAX3* (55% ARMS) or *PAX7* (23% ARMS) respectively, leading to myogenic proliferation and dedifferentiation through increased PAX activity. Twenty-five per cent ARMS is fusion-negative and has similar outcomes to ERMS. *PAX3–FOXO1* fusion ARMS has a poorer prognosis than *PAX7–FOXO1* ARMS. Classification of RMS will be based on these gene fusions rather than their histology in future Children's Oncology Group (COG) studies.

Pleomorphic (anaplastic) rhabdomyosarcoma

Histologically, pleomorphic RMS appears as sheets of large polygonal multinucleated eosinophilic cells that can be round or spindle-shaped. It has no recurrent genetic structural rearrangements although *10q23* loss and *1p22–23* gains are typical. This rare and aggressive tumor of childhood typically arises in the lower limbs, grows rapidly, and is often painful.

Spindle-cell/sclerosing rhabdomyosarcoma

Histologically, spindle-cell RMS has a fascicular pattern with rare rhabdomyoblasts and occasional cross-striations. The immunohistochemical staining pattern is similar to that of ERMS with limited myogenin expression and strong *myoD1* positivity. Paratesticular RMS is usually spindle cell.

MULTIMODAL THERAPY

Multimodal therapy is the mainstay of RMS management and combines surgery, RT, and chemotherapy. Surgical resection should precede other modalities if negative margins are achievable while avoiding disfigurement and loss of function. For the majority of patients who present with unresectable disease, treatment strategies differ depending on the protocols used, however the general principles are to reduce the tumor in size with neoadjuvant chemotherapy to the point where local therapy — either surgery and/or RT — will achieve local control.

Children's Oncology Group (COG) versus Société Internationale d'Oncologie Pédiatrique (SIOP)

There is a difference in approach between the two main pediatric oncological cooperative groups. The approach of the COG is to use chemotherapy in order to render the tumor more resectable and then to use RT in patients with micro- or macroscopic residual disease in order to avoid relapse and consequent salvage therapy. SIOP's approach is to use aggressive systemic therapy to reduce the use of local therapies and avoid the complications associated with them. Failures are treated with surgery in preference to RT. Long-term morbidity data support SIOP's approach and survival data support the COG approach.

Chemotherapy

The backbone of chemotherapy for RMS is a combination of vincristine, actinomycin-D, and cyclophosphamide (VAC). A subset of patients with low-risk RMS can be treated with vincristine and actinomycin-D (VA) alone. Little progress in outcome or dose reduction has been made with high-risk RMS, which is currently treated with VAC alternating with vincristine and irinotecan (VAC/VI).

Radiotherapy

RT can be used as primary local therapy and is essential in the adjuvant setting for the prevention of local recurrence in patients with gross or microscopic residual disease. Only Group I patients with ERMS are managed without RT. Regimens depend on the site of the tumor, histology, amount of residual disease, and presence of involved lymph nodes. Three-dimensional computer planning and techniques that spare normal tissue should be used such as proton beam therapy or intensity-modulated radiation therapy (IMRT). Brachytherapy has been used effectively for RMS of the genitourinary tract, head and neck, and in patients less than 3 years of age. If required, even in young children, RT should not be avoided nor the dose reduced in violation of protocol as it leads to an increased risk of local failure.

SURGERY

Anesthesia

Most procedures are conducted under general anesthesia. Multiple procedures may be carried out under the same anesthetic in order to minimize the number given.

Biopsy

Imaging for staging should precede biopsy in order to determine the best anatomical approach to the tumor, suggest which area is best representative of the underlying disease, and avoid radiological artifact at the biopsy site that may affect the interpretation of the images. Enough tissue for pathological, biological, cytogenetic, and treatment protocol studies should be procured. Incisional, excisional, and core-needle biopsies are acceptable depending on the circumstances. Fine-needle aspiration biopsy should not be used as the samples are inadequate for the evaluation of tissue architecture. Open, laparoscopic, or thoracoscopic techniques can be used for intra-abdominal or intrathoracic primary or metastatic disease. Core-needle biopsy is less invasive and is being successfully used in most anatomic locations, though it has a slightly higher rate of diagnostic failure. Multiple cores are required for diagnosis. Unplanned incisional or needle biopsies may complicate surgical excision and compromise the preservation of cosmesis and function (**Table 61.4**: Procedure 1).

Table 61.4 **Key operative steps in RMS procedures**

Procedure 1: Incisional/core biopsy

Patient position	Supine – anterior tumors Lateral – lateral and posterior tumors
Incision/needle-track	Plan the shortest route from the skin to the lesion It should be easily resectable en bloc with the specimen Do not violate more than one anatomical compartment It should be remote from the main neurovascular bundle to that area
Tips and tricks for incisional biopsy	Use the smallest longitudinal incision required to obtain a representative sample • Limbs – along the long axis • Chest wall – in line with the ribs Use a scalpel rather than curettage or cautery in order to preserve tissue architecture Avoid hematomas as they complicate surgical excision by making tumor margins indistinct Use drains that exit close to and in line with the incision – the drain track is contaminated and must be resected with the specimen
Tips and tricks for core-needle biopsy	Use a 14- or 16-gauge tru-cut needle that provides a 20 mm long sample Use image (USS or CT) guidance if the lesion is not palpable Take multiple samples

Procedure 2: Extremity RMS resection

Patient position	Supine – anterior tumors (e.g. medial arm, dorsal forearm) Lateral – lateral tumors (e.g. hip, lateral or medial thigh) “Frog-legged” – medial thigh tumors Prone – posterior tumors Arm table – useful for upper limb tumors
Markings	Mark the skin to be resected (including the biopsy site) with at least a 1 cm margin Mark the limit of the skin flaps Mark important anatomical structures Inguinal resection – femoral neurovascular bundle Cubital fossa – brachial artery, median, ulnar and radial nerves
“Prep and drape”	Use a “limb shut-off” method Use a tourniquet if required
Resection	Incise and raise skin flaps medially and laterally beyond the peripheral margins of the tumor Tumor should be covered with a margin of at least 0.5–1.0 cm of normal tissue, or an intact uninvolved fascial, adventitial, epineurial, or periosteal layer Apply titanium clips to close margins in the tumor bed to direct adjuvant radiotherapy

Table 61.4 **Key operative steps in RMS procedures**

Pathology	Send a frozen section of any margin in doubt Deliver the resected specimen to the pathologist for an in-person discussion regarding specimen orientation, margins, and peritumoral anatomy
Reconstruction	As per the plastic and reconstructive surgical team
Procedure 3: Chest wall RMS resection	
Position	Supine – sternal and parasternal tumors Lateral – anterolateral, lateral, or posterolateral tumors If in the lateral position, the lumbar spine should be over the mid-table break so that lateral flexion can be achieved Place an axillary roll to protect their brachial plexus Slight hip and knee flexion with a pillow between the legs Shoulders in ~90° flexion with the upper arm supported
Markings	Mark the skin to be resected (including the biopsy site) with at least a 1 cm margin Mark the limit of the skin flaps Mark important anatomical structures (e.g. spine, scapula, and ribs)
“Prep and drape” Resection	Beyond the midline anteriorly and posteriorly to the opposite side of the lesion Incise along or about the rib of interest and raise skin flaps superiorly and inferiorly beyond the margins of the tumor Involved chest wall muscle (e.g. latissimus dorsi and/or the serratus anterior) should be resected otherwise a muscle sparing approach is recommended
Resection	Divide the involved segment of rib/s and intercostal muscles anteriorly and posteriorly at least 2 cm beyond the tumor Resect the periosteum of the uninvolved rib above and below the tumor Involved lung should resected en bloc with a linear stapler or by formal lobectomy Inspect the pleural and remove any suspicious lesions Resect involved diaphragm en bloc. Care should be taken not to contaminate the abdominal cavity with tumor if the diaphragmatic peritoneum is breached Apply titanium clips to close margins in the tumor bed to direct adjuvant radiotherapy
Pathology	Send a frozen section of any margin in doubt Deliver the resected specimen to the pathologist for an in-person discussion regarding specimen orientation, margins, and peritumoral anatomy
Reconstruction	As per the plastic and reconstructive surgical team
Procedure 4: Inguinal orchiectomy for paratesticular RMS resection	
Position	Supine

Table 61.4 **Key operative steps in RMS procedures**

Exposure and tumor isolation	Oblique inguinal incision to allow extension for larger masses Open the anterior wall of the inguinal canal through the external inguinal ring Atraumatically control the spermatic cord at the deep inguinal ring Deliver the testis into the wound Isolate the testis from the operative field in a non-permeable bag Divide the spermatic cord at the deep inguinal ring
Frozen sections	Diagnostic incisional biopsy for frozen section – only if the diagnosis is unclear Send a frozen section of any margin in doubt (e.g. proximal spermatic cord)
Closure	Close in layers according to surgeon preference

Procedure 5: Sentinel lymph node biopsy

Lymphoscintigraphy	4–6 hours preoperatively – intradermal injection of 0.1 mL 125 uCi of ^{99m}Tc-labeled sulfur colloid, ^{99m}Tc human serum albumin, ^{99m}Tc albumin colloid or ^{99m}Tc-labeled albumin non-colloid at four peritumoral sites Large field view camera and high resolution collimator imaging CT co-localization of sentinel nodes Image review by surgeon and nuclear medicine physician
Intraoperative technique	General anesthetic Position for easy access to the tumor • Supine – anterior tumors • Lateral – lateral and posterior tumors Inject 3 mL of 1% vital blue dye (isosulfan blue or patent blue) intradermally at four peritumoral sites Reposition the patient for access to the lymph node site of interest • Supine – cervical, axillary and inguinal nodes • Lateral – popliteal and scapular nodes Prep and drape the patient Locate the sentinel node using the lymphoscintographic images and a gamma probe Mark and incise with a potential future lymph node dissection in mind (e.g. transverse in the axilla; longitudinal in the groin) Localize the sentinel node/s using the gamma probe and visualization of blue lymphatics Excise all blue-stained and/or lymph nodes with activity exceeding 10% of that of the most radioactive node If the background radiation is <10% of the most radioactive node, more sentinel nodes are present in the operative field and should be found and removed Drains are not usually required Closure in layers according to surgeon preference

Table 61.4 Key operative steps in RMS procedures

Pathology	Label all nodes and send the specimen fresh Direct communication with the pathologist is essential Slides should be sectioned and analyzed in 50–100 μm wide slices

Surgical principles of RMS resection

Surgical resection of RMS is site-specific. It should only be attempted if a complete resection is anticipated with a 0.5–1.0 cm margin of healthy tissue or an uninvolved fascial or periosteal margin. Exceptions to wide margins apply if they are technically impossible or if they lead to unacceptable loss of cosmesis or function. Preoperative planning should include discussions with the involved medical and radiation oncologist, radiologist, and pathologist at a tumor board so that surgery can be coordinated with the other treatment modalities. Following excision, the tumor should be accurately oriented and the tumor bed marked with titanium clips in order to guide further therapy if the excision is incomplete or margins are inadequate. If narrow margins are unavoidable, frozen sections of tissue from the margins should be taken intraoperatively for evaluation. The tumor should not be bisected by the surgeon prior to pathological evaluation. In the event of a non-oncological resection or if macro- or microscopic margins are positive or unclear, primary re-excision (PRE) is recommended in order to achieve negative margins.

If primary excision or PRE is not possible, neoadjuvant chemotherapy should commence with re-evaluation at approximately 12 weeks. If possible, delayed primary excision (DPE) should be carried out, followed by adjuvant RT. Those with unresectable disease after neoadjuvant chemotherapy should receive neoadjuvant RT followed by DPE. DPE is preferred to primary RT and allows for adjuvant RT dose reduction in both low- and intermediate-risk disease. If the tumor remains unresectable, higher-dose primary RT should be considered, after which surgery may or may not be required. At this point, if the tumor remains uncontrolled, definitive mutilating surgery can be considered. These surgeries are defined as orbital exenteration, major facial resection, bladder or prostatic resection, hysterectomy, pelvic exenteration, urinary diversion, pneumonectomy, and limb amputation. The outcomes of tumor debulking are no better than biopsy alone. In some circumstances debulking worsens local recurrence and it is not recommended.

Second-look operations

Following the completion of primary therapy, reimaging is performed with CT or MRI per protocol. Persistent disease, partial response, non-response, or uncertainty regarding the outcome of primary therapy is an indication for a second-look operation (SLO). This is most useful for truncal and extremity tumors. The objective of SLO is to evaluate response, re-biopsy residual tumor, and complete the resection in order to achieve local control. Approximately 12% of complete responders harbor tumor on SLO but many of these are resectable. In 75% of patients, SLO leads to reclassification of partial responders to complete responders following resection of residual tumor.

SITE-SPECIFIC OPERATIONS

Head and neck

Because of the difficulty of achieving adequate margins without significant cosmetic and functional compromise, operative intervention is generally limited to biopsy alone followed by chemotherapy and RT. Cervical lymph node biopsy is not required unless there is clinical nodal involvement.

ORBITAL RMS

Orbital tumors should be biopsied with the help of an ophthalmologist. Orbital exenteration is reserved for the few patients who have refractory or recurrent disease. In the interests of preserving vision, RT should be limited to the conjunctiva, cornea, and lens. Survival is 90–95%.

NON-ORBITAL AND CRANIAL PARAMENINGEAL RMS

These include tumors of the middle ear, mastoid, nasopharynx, nasal cavity, paranasal sinus, parapharyngeal, pterygopalatine, and infratemporal fossae. Those without involvement of the cerebrospinal fluid do not require whole brain irradiation. Sentinel lymph node biopsy may be beneficial for staging. Craniofacial resection should only occur if primary management fails.

NON-ORBITAL AND NON-PARAMENINGEAL RMS

These include tumors of the oral cavity, larynx, parotid, cheek, and scalp, and soft tissue of the neck. Provided cosmesis and function are not compromised, wide excision with ipsilateral neck node sampling of clinically involved nodes is indicated. For unresectable tumors, operative management should be considered if other treatment modalities fail.

Complex resection of head and neck RMS is beyond the scope of this chapter and should be done with the help of specialist surgical teams. These may include head and neck surgeons, maxillofacial surgeons, ophthalmologists, neurosurgeons, skull-base surgeons, and plastic and reconstructive surgeons.

Extremities

Limb-sparing resection should follow neoadjuvant therapy (**Table 61.4**: Procedure 2). Muscle resection from origin to insertion is no longer recommended. Reconstruction should be planned preoperatively with a plastic and reconstructive surgical and/or orthopedic team and may include native or donated arterial or venous grafts with or without synthetic or arterial or venous patches, primary nerve repair, nerve transfers or grafts, skin grafts (split or full-thickness) or local flaps, and pedicled or free myocutaneous flaps. For young children, reconstruction must take their projected growth into account. Amputation is rarely required, does not improve overall survival as a primary procedure, and is reserved for persistent or recurrent disease not amenable to limb preservation. RMS of the hands and feet is treated with primary chemotherapy and RT, with amputation reserved for persistent or recurrent disease.

Regional lymph node involvement in extremity and truncal RMS affects 30–40% of patients and is a poor prognostic indicator. All clinically suspicious lymph nodes should be biopsied. Lymphatic mapping and sentinel lymph node biopsy is indicated in patients with clinically and radiologically node negative disease (**Table 61.4:** Procedure 5). Positive nodal basins should be treated with RT followed by SLO and surgical resection in the event of treatment failure.

Trunk

ABDOMINAL WALL RMS

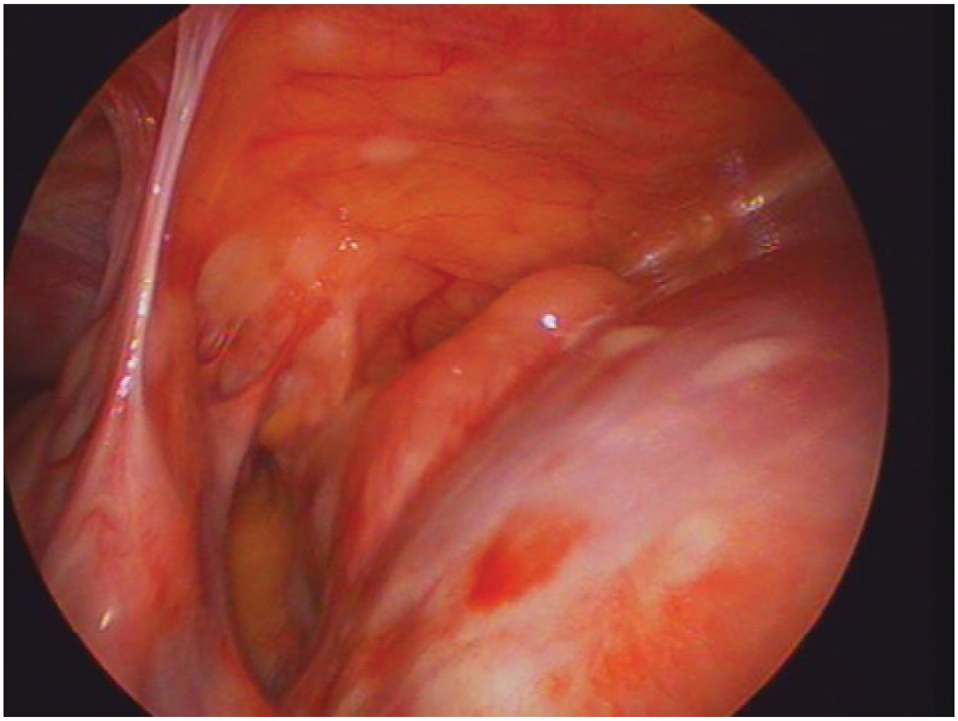
The resection of abdominal wall tumors usually necessitates a full-thickness excision from the skin to the peritoneum. If tumor invades the abdominal cavity and involves intra-abdominal organs, they should be resected with the abdominal wall en bloc.

ABDOMINAL WALL RECONSTRUCTION

Abdominal wall reconstruction is important in order to avoid an incisional hernia. Myofascial replacement can be achieved with mesh (biological or synthetic) and/or native tissue transfer by abdominal component separation, myocutaneous pedicled (e.g. anterolateral thigh flap based on the medial circumflex femoral artery) or free flaps. The spectrum of available meshes, component separation techniques, and various flaps is beyond the scope of this chapter and quality evidence supporting one over the other is scant. Growth in small children is less likely to be impacted by the type of abdominal reconstruction used. Preoperative consultation with a plastic and reconstructive surgeon is recommended.

CHEST WALL RMS

Prior to excision, thoracoscopy may assist in evaluation of intrathoracic disease and lung involvement (**Figure 61.2**). Definitive chest wall excisions often involve the skin (including the biopsy site), underlying subcutaneous tissue and muscle, involved ribs and pleura, the periosteum of the rib above and below the lesion, and, frequently, a wedge of underlying lung and part of the diaphragm (**Table 61.4:** Procedure 3). The specimen should be removed en bloc. Normal ribs should not be removed. A two or three rib resection is acceptable in a prepubertal child and avoids growth-related scoliosis. A four or five rib resection to achieve R0 is an unnecessary cosmetic and functional sacrifice. Adjuvant RT provides adequate local control and there is no difference in overall survival in chest wall RMS between those patients who have had an R0 and an R1 resection. Clinically positive nodes should be biopsied. If none is evident, a sentinel lymph node biopsy is indicated (Table 61.4: Procedure 5).



61.2 Thoracoscopy demonstrating malignant adhesions between the lung and chest wall and rhabdomyosarcomatous deposits on the lung, chest wall, and hemidiaphragm.

CHEST WALL RECONSTRUCTION

The objectives of chest wall reconstruction include the restoration of chest wall rigidity and protection of the underlying organs, prevention of pulmonary or cardiac herniation, preservation of respiratory function, avoidance of scapular trapping, and an acceptable cosmetic outcome. The indications for reconstruction include resection of four or more ribs and lateral, anterolateral, costal, and sternal resections. Resections beneath the scapula or pectoral muscles and posterior and paraspinal resections do not require reconstruction unless there is a risk for scapula trapping on adduction of the shoulder from an abducted position.

Chest wall reconstruction should be done with the help of a plastic and reconstructive surgical team and can involve rib grafts or implants, mesh, and/or pedicled or free myocutaneous flaps. Consideration for future growth of the patient is a major issue.

PARASPINAL, RETROPERITONEAL, AND PELVIC RMS

These are usually unresectable due to their size, invasiveness, and proximity to vital structures (e.g. spinal canal and major vessels). Neoadjuvant chemotherapy +/− RT and DPE is the usual approach. Paraspinal and pelvic bony resections require the input of specialist neurosurgical and orthopedic teams. Limited resections not involving the hip or sacroiliac joint are tolerated well with preservation of the patient's ability to ambulate. Without adjuvant RT, recurrence is probable following R1 or R2 resections.

INTRA-ABDOMINAL RMS

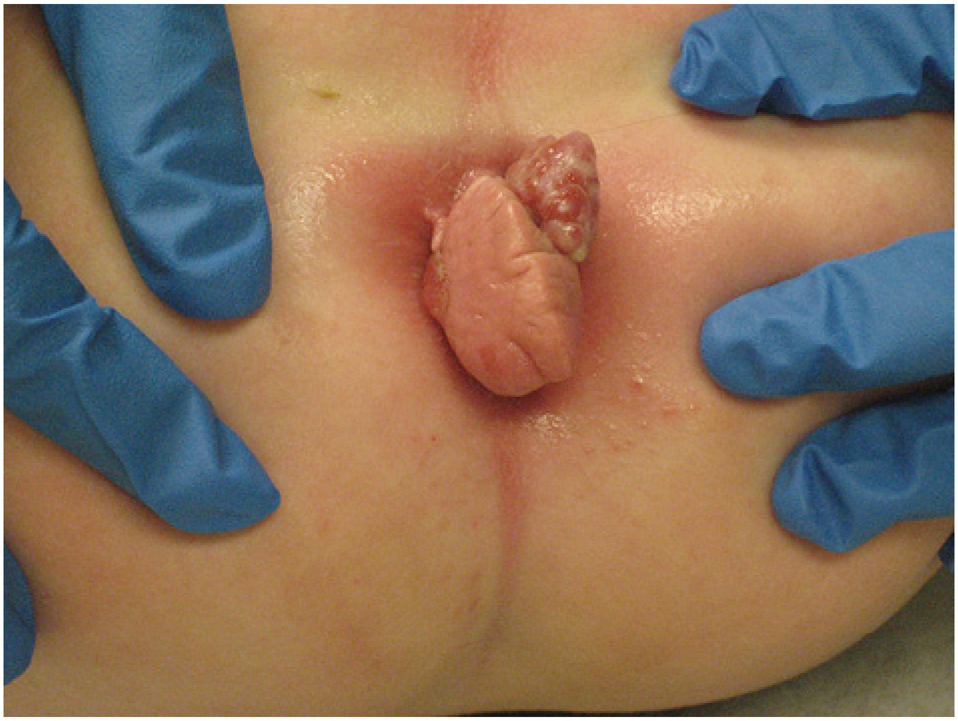
The technical details for the operative resection of intra-abdominal RMS are as diverse as the range of organs in which it appears and may require formal hepatic segmentectomy or lobectomy, splenectomy, bowel resections, and en bloc abdominal wall and/or diaphragmatic resections while avoiding functional compromise (e.g. liver failure or short gut). The local control of peritoneal sarcomatosis may require cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC).

BILIARY RMS

Biliary RMS has an excellent prognosis. Most is botryoidal and can be managed with chemotherapy and RT. Primary excision or PRE is rarely necessary. Biliary obstruction usually resolves once chemotherapy commences and instrumentation of the biliary tree in the absence of sepsis may increase the incidence of infection. Liver transplant for unresectable biliary RMS without extrahepatic metastases has been reported in children but there are little long-term outcome data. A detailed operative description is beyond the scope of this chapter.

PERINEAL AND PERIANAL RMS

Perineal and perianal RMS (**Figure 61.3**) can be mistaken for perianal abscess, is usually unresectable, and confers a poor prognosis. Following biopsy, neoadjuvant chemotherapy followed by DPE and RT is recommended. Fecal diversion should be considered if there is anorectal obstruction. Clinically suspicious lymph nodes should be biopsied. More aggressive surgery including sphincter sacrifice should be reserved for failure of primary therapy. The detailed technical aspects of perineal and perianal resections, diverting sigmoid colostomy, permanent sigmoid colostomy, resection of perianal tumors, and abdominoperineal resection are beyond the scope of this chapter.



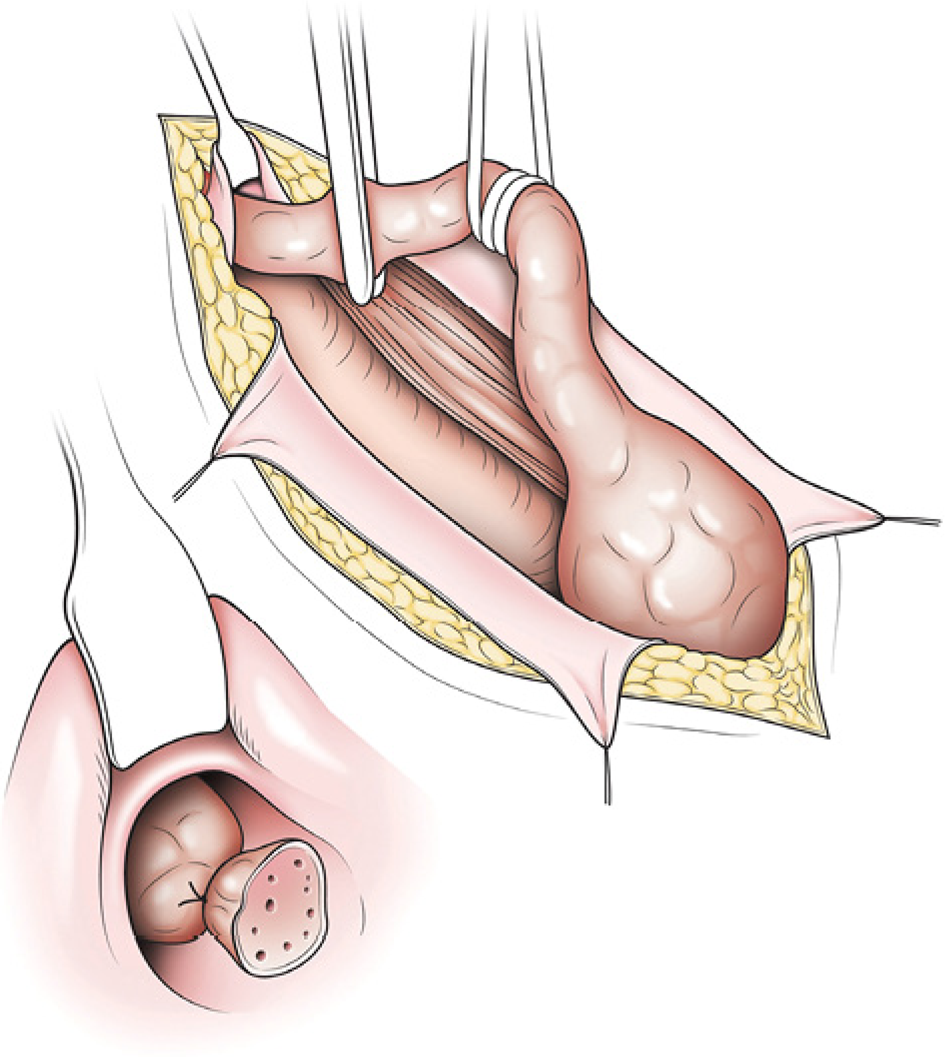
61.3 Perianal rhabdomyosarcoma in an infant initially diagnosed with condylomata.

Genitourinary tract

Rhabdomyosarcoma is the most common pediatric malignancy of the genitourinary tract.

PARATESTICULAR RMS

Paratesticular RMS originates from the epididymis, spermatic cord, testis, and tunica albuginea, is usually spindle cell, and usually presents early as a painless scrotal mass. It is usually primarily resectable by inguinal orchiectomy (**Figure 61.4**) with high ligation and complete resection of the spermatic cord and has an excellent prognosis (**Table 61.4**: Procedure 4). If the tumor is biopsied via the scrotum or fixed to it, or if there is tumor spillage, hemiscrotectomy or hemiscrotal RT is recommended in order to avoid recurrence. The contralateral testis should be transposed out of the field and into the abdomen or a thigh pouch to preserve fertility.



61.4 Right inguinal orchiectomy for paratesticular rhabdomyosarcoma.

All patients over 10 years of age and any patient with radiologically suspicious nodes should undergo retroperitoneal lymph node sampling. Positive retroperitoneal nodes are treated with formal ipsilateral retroperitoneal lymph node dissection and/or RT. Complications include lymphocele, ejaculatory dysfunction, lymphedema of the lower extremity, and intestinal obstruction.

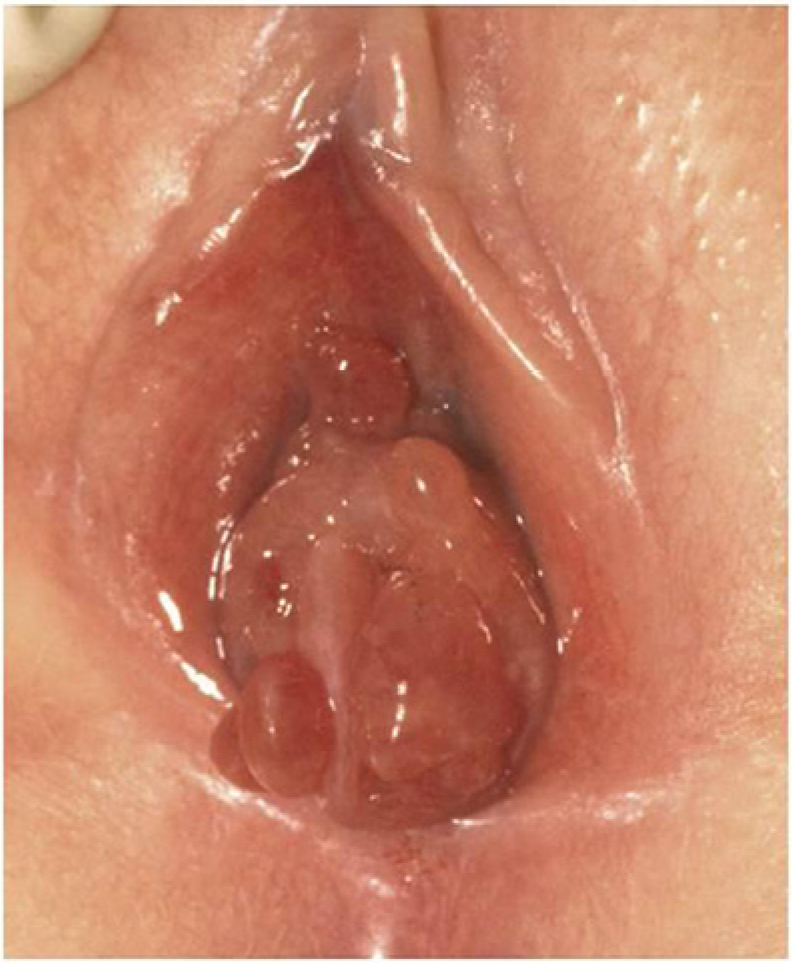
BLADDER/PROSTATE RMS

Bladder and prostate RMS is usually unresectable at diagnosis. Biopsy can be cystoscopic, via a transabdominal open or laparoscopic approach, or via a core transrectal USS or transabdominal CT-guided approach. Preservation of bladder function is paramount. Uncommonly, small tumors of the bladder dome are primarily resectable. More commonly, management is with neoadjuvant chemotherapy +/- RT followed by DPE. More extensive, particularly distal bladder resections may require ureteric reimplantation and/or bladder augmentation. Bladder outlet obstruction should be managed with urethral catheterization. If suprapubic catheterization is required, the track will need excision due to the risk of seeding.

In prostatic RMS, omission of adjuvant RT or the use of neoadjuvant brachytherapy followed by surgical resection helps to preserve bladder function. Retroperitoneal lymph node sampling of clinically or radiologically suspicious nodes is recommended. Positive nodes are treated with RT and/or surgery. Biopsy proven residual or persistent prostatic disease is managed with limited resection and brachytherapy. Radical surgery is a last resort. This may include partial or total cystectomy with or without bladder augmentation, ureteric reimplantation, radical prostatectomy, urethral reconstruction, and/or pelvic exenteration with continent urinary diversion, the details of which are beyond the scope of this chapter.

VULVA/VAGINA/UTERUS RMS

Vaginal, vulval, and uterine rhabdomyosarcoma are usually botryoidal and present with vaginal discharge, bleeding, or prolapse of a polypoid mass (**Figure 61.5**), which may be primarily resectable. Diagnosis is made by vaginoscopic or open incisional or excisional biopsy. Eighty-five per cent of patients require biopsy only followed by chemotherapy and RT or brachytherapy, with little if any surgical intervention and excellent outcomes (5-year FFS 92%). If surgery is required, an R1 resection with adjuvant RT is preferable to radical surgery. Hysterectomy, vulvectomy, partial vaginectomy, or sleeve resection are only indicated for persistent or recurrent disease. Bladder salvage is almost always possible even with locally advanced tumors. Oophorectomy should only be considered if there is direct ovarian involvement. Pelvic and retroperitoneal lymphatic metastases are uncommon and lymph node evaluation is not routinely performed. The details of hysterectomy, sleeve-vaginectomy, vulvectomy, and oophorectomy are beyond the scope of this chapter.

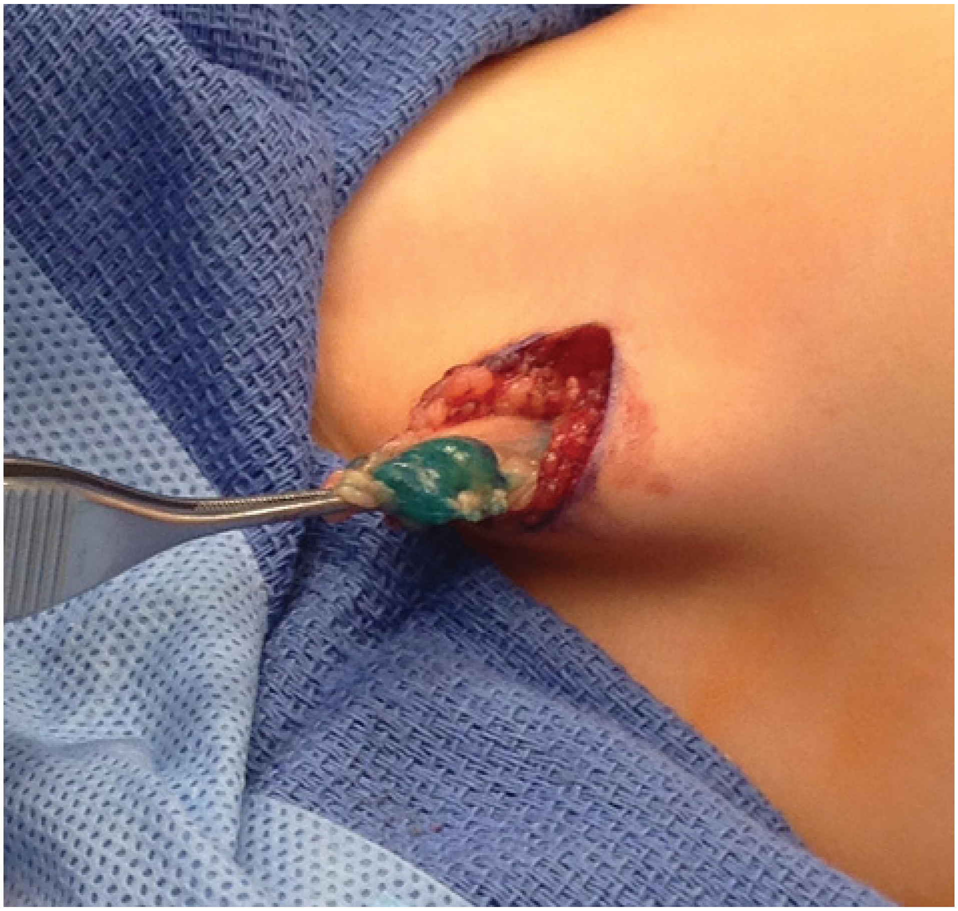


61.5 Botryoidal (grape-like) vaginal rhabdomyosarcoma in an infant.

To preserve fertility, oophoropexy or ovarian tissue preservation should be considered prior to the commencement of RT. Oophoropexy can be done via laparotomy or laparoscopy and can be medial and behind the uterus, lateral and out of the radiation field, subcutaneous in the abdominal or flank wall, or using heterotopic ovarian transplantation. Exposure to alkylating agents such as cyclophosphamide at doses >4.8 mg/m^2 predict ovarian failure irrespective of radiation exposure and may negate the need for transposition.

Lymph node evaluation and sentinel node biopsy

Lymph node positivity is an independent risk factor for poor prognosis. Clinically suspicious lymph nodes should be biopsied (**Table 61.4:** Procedure 5). Patients with extremity and truncal RMS with clinically negative nodes should undergo sentinel node biopsy which allows for evaluation of nodal status without the morbidity associated with complete nodal dissection. Nodal positivity is an indication for management with RT with surgery reserved for therapeutic failures (**Figure 61.6**).



61.6 Operative photograph demonstrating the excision of axillary sentinel lymph node stained with patent blue dye.

RECURRENT RHABDOMYOSARCOMA

Risk factors for local recurrence include age >10 years, tumor >5 cm, unfavorable site, alveolar histology, “fusion-positivity,” lymph node/s positivity, and metastatic disease at diagnosis. Short time to recurrence is a poor prognostic indicator. There is no standard treatment for recurrent RMS. In general, chemotherapy is initiated to establish a response followed by local control with surgery and/or primary RT.

METASTATIC RHABDOMYOSARCOMA

Approximately 15% of patients present with metastatic (Group IV) disease. The outcome for these patients remains poor. Improved prognosis is associated with two or fewer metastatic sites, embryonal histopathology, age 1–9 years, favorable site, and uninvolved bone marrow. Most metastatic disease is treated non-operatively. Surgery is recommended for biopsy to confirm the diagnosis and occasionally for removal of metastatic disease that has persisted after chemotherapy. Primary site resection and RT may improve outcome even in the setting of metastatic disease. A number of patients with peritoneal rhabdosarcomatosis have been treated with CRS and cisplatin-based HIPEC, however results have not been conclusively beneficial and trials are in progress with other agents including doxorubicin.

COMPLICATIONS

Complications include wound infection and postoperative bleeding, which should occur in <5% patients. Seroma may occur and should be aspirated if symptomatic. Following sentinel lymph node biopsy in the inguinum or axilla, lymphedema occurs in ~5% patients but is not usually clinically signficant. Occasionally, it needs to be addressed with compression garments, physiotherapy, and skin care.

FURTHER READING

Austin, MT, Andrassy R. Soft tissue sarcoma. In: Carachi R, Grosfeld JL (eds). *The Surgery of Childhood Tumours*, 3rd ed. Berlin: Springer; 2015: pp. 345–81.

Dasgupta R, Fuchs J, Rodeberg D. Rhabdomyosarcoma. *Semin Pediatr Surg* 2016; 25(5): 276–83.

Ferrari A, Miceli R, Meazza C, et al. Soft tissue sarcomas of childhood and adolescence: The prognostic role of tumor size in relation to patient body size. *J Clin Oncol* 2009; 27(3): 371–6.

Harrison D, Parisi MT, Shulkin BL, et al. 18F 2fluoro-2deoxy-D-glucose positron emission tomography (PET-FDG) response to predict event-free survival (EFS) in intermediate risk (IR) or high risk (HR) rhabdomyosarcoma (RMS): A report from the soft tissue sarcoma committee of the children's oncology group (COG). *J Clin Oncol* 2016; 34(15 suppl): 10549.

Hayes-Jordan A, Doherty DK, West SD, et al. Outcome after surgical resection of recurrent rhabdomyosarcoma. *J Pediatr Surg* 2006; 41(4): 633–8.

Hayes-Jordan A, Green H, Lin H, et al. Cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) for children, adolescents, and young adults: The first 50 cases. *Ann Surg Oncol* 2015; 22(5): 1726–32.

Khullar OV, Fernandez FG. Prosthetic reconstruction of the chest wall. *Thorac Surg Clin* 2017; 27(2): 201–8.

Parham DM, Montgomery EA, Nascimento AF, Barr FG. Skeletal–muscle tumors. In: Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F (eds). *WHO Classification of Tumors of Soft Tissue and Bone*, 4th ed. Lyon: International Agency for Research on Cancer; 2013: pp. 123–35.

PDQ® Pediatric Treatment Editorial Board. *PDQ Childhood Rhabdomyosarcoma Treatment*. Bethesda, MD: National Cancer Institute. Updated 7 May 2020. Available at: https://www.cancer.gov/types/soft-tissue-sarcoma/hp/rhabdomyosarcoma-treatment-pdq [Accessed 1 June 2020.]

Rodeberg DA, Paidas CN, Lobe TL, et al. Surgical principles for children/adolescents with newly diagnosed rhabdomyosarcoma: A report from the Soft Tissue Sarcoma Committee of the Children's Oncology Group. *Sarcoma* 2002; 6(4): 111–22.

62

Sacrococcygeal teratoma

AGOSTINO PIERRO, MIGUEL GUELFLAND, AND ANNIKA MUTANEN

INTRODUCTION

Sacrococcygeal teratoma (SCT) was first described by Virchow in 1869, its name being derived from the Greek word meaning “monstrous tumor.” They represent the most common germ-cell tumors of childhood (40%), with an incidence of about 1 in 40 000 live-births, and a female preponderance of 4 : 1.

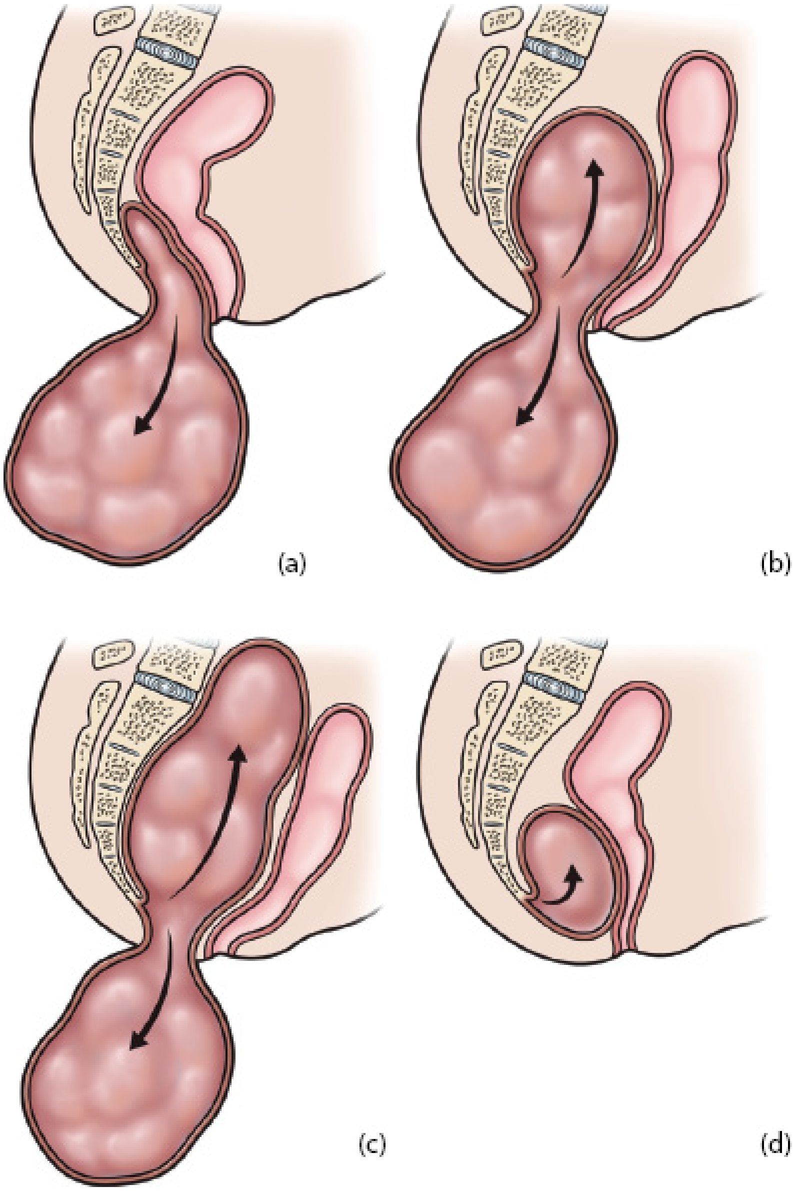
PATHOLOGY

Embryologically, SCTs arise from totipotent primordial endodermal germ cells, but they may contain tissues originating from all three germ layers, including dermal elements (e.g. skin), muscle, glial tissue, intestinal mucosal, and pancreas. Histologically, SCT can be classified as mature (~70%), immature with embryonic components (~20%), or immature with malignant components (~10%).

SCTs are classified according to anatomic criteria first established by the Surgical Section of the American Academy of Pediatrics.

- *Type I (~50%):* Pedunculated tumor predominantly external with minimal extension into the presacral region (**Figure 62.1a**)
- *Type II (~30%):* More significant intrapelvic extension (**Figure 62.1b**)

- *Type III (~10%):* Predominantly pelvic tumor with abdominal extension, but minimal external component (**Figure 62.1c**)
- *Type IV (10%):* A completely internal presacral tumor without external evidence of disease (**Figure 62.1d**)



62.1a–d Classification of SCT. (a) Type I, (b) Type II, (c) Type III, and (d) Type IV.

Malignant SCTs almost exclusively arise from embryonal carcinomas or yolk-sac tumors and are more commonly seen in recurrent lesions and in those with incomplete resection.

- >90% are benign at birth, but risk of malignancy increases with age at diagnosis:
  - Up to 50% malignancy at 6 months
  - Up to 75% at 1 year
- According to a study by Altman et al., the risk of malignancy increases with more hidden lesions:
  - 8% in type I
  - 21% in type II
  - 34% in type III
  - 38% in type IV

STAGING

SCTs are currently staged using The Children's Cancer Study Group and Pediatric Oncology Group staging system for extragonadal germ-cell tumors (**Table 62.1**).

Table 62.1 **Staging system used for SCT**

Stage	Extent of disease
I	Complete excision with coccygectomy Negative tumor margins Tumor markers positive, but fall to normal if negative at diagnosis Lymphadenectomy must be negative for tumor
II	Microscopic residual tumor; lymph node negative Tumor markers positive or negative
III	Gross residual tumor Retroperitoneal nodes negative or positive; tumor markers positive or negative
IV	Distant metastases, including liver

DIAGNOSIS

Prenatal diagnosis and management

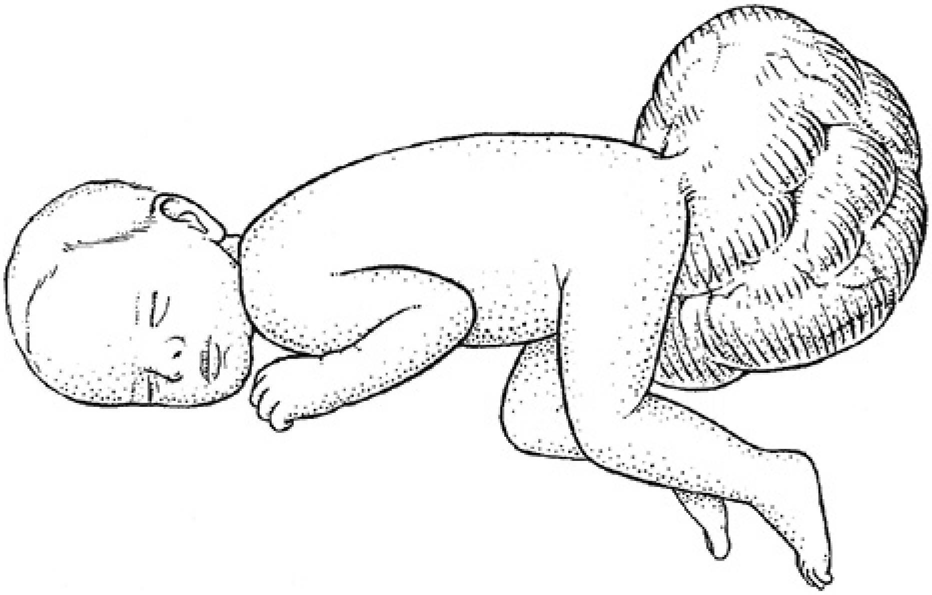
Routine use of serial prenatal ultrasound (US) has increased the detection of SCT and allowed for accurate identification of site and details of intrapelvic extension or urinary tract obstruction. The combination of conventional US, color-flow Doppler, and echocardiography is essential for effective perinatal management. Fetal magnetic resonance imaging (MRI) also helps to define the anatomy of the tumor accurately.

Caesarean section is advocated for tumors >5 cm, as dystocia during vaginal delivery may cause tumor rupture, avulsion, or hemorrhage, and can be fatal. Poor prognostic factors during fetal life include polyhydramnios, placentomegaly, and hydrops fetalis. Recent-onset polyhydramnios may also be associated with preterm labor. Perhaps up to 20% of all SCT detected prenatally develop hydrops from high-output cardiac failure secondary to arteriovenous shunting within the tumor, which can be fatal.

Fetal interventions such as amniodrainage, cyst aspiration, and relief of bladder outflow obstruction can be performed in those cases where hydrops and prematurity are present. Consideration of open fetal surgery for debulking of SCTs is only considered in very select cases.

Diagnosis in the neonate and children

Most SCTs present with a large skin-covered mass protruding externally from the coccygeal region and are therefore diagnosed clinically at birth (**Figure 62.2**).



62.2

There may be large vessels or ulcers visible on the tumor surface. In all cases, the tumor is attached to the coccyx (therefore the necessity of coccygectomy during tumor excision) and projects to a varying degree into the presacral space between the sacrum and the rectum. In most cases, the blood supply to the tumor is derived from the middle sacral artery. Although most neonates with SCT are asymptomatic, this upward extension into the pelvic space may compress and elevate the rectum, vagina, bladder, and uterus, causing symptoms of constipation, large bowel obstruction, urinary retention, an abdominal mass, or symptoms of malignancy, such as failure to thrive.

Most SCTs are isolated though associated anomalies are identified in up to 20% of infants, including central nervous system (25%, e.g. anencephaly, trigonocephaly, Dandy–Walker malformation, spina bifida, and myelomeningocele); musculoskeletal system (25%, e.g. congenital dislocation of hips, vertebral anomalies, and the Currarino triad of anorectal malformation, sacrococcygeal bony defect, and presacral mass); and urogenital anomalies (hypospadias, vesicoureteric reflux), or duplications of the vagina or uterus.

Differential diagnosis

The major differential diagnosis is with a myelomeningocele. Clinically, SCTs are a more solid presacral tumor while anterior meningoceles are obviously cystic, may have an anterior sacral defect, and pressure on the sacral mass will result in a bulging fontanelle. Less common differentials include lipomeningocele, hemangioma, lymphangioma, chordoma (50% of these occur in the sacrococcygeal region), pelvic neuroblastoma, sarcoma, hamartoma, cystic duplication of the rectum, neuroenteric cysts, dermoid cysts, meconium pseudocysts, and perirectal abscess. The differential diagnosis is confirmed by radiography, including US, computed tomography (CT), or MRI.

PREOPERATIVE INVESTIGATIONS

Plain radiographs may show calcifications in the tumor (up to 60%) or identify spinal defects. US, MRI, or CT scans determine the level of intra-abdominal or intrapelvic extension. Markers such as α-fetoprotein (AFP) and β-human chorionic gonadotropin (β-hCG) may be useful for the assessment of disease progression but at birth needs to be interpreted carefully. Serum AFP is normally elevated hugely at birth and remains above normally quoted levels for up to 6 months. However, most yolk-sac tumors and some embryonal carcinomas also secrete AFP. Since malignant elements of SCT almost exclusively arise from one of these two sources, AFP measurement can be a useful marker for malignant degeneration of benign lesions, or for the presence of residual or recurrent malignant disease. It can be measured in the serum and demonstrated in the cells by immunohistochemistry. Persistently high levels may be an indication for further surgery or chemotherapy. Serum β-hCG and, rarely, carcinoembryonic antigen may also be elevated and can be produced by choriocarcinomas.

PRENATAL MANAGEMENT

Management is based on fetal lung maturity and the presence or absence of placentomegaly and hydrops. Early elective delivery by Caesarean section is indicated upon reaching fetal lung maturity in the presence of evidence of fetal (and maternal) distress such as hydrops. Fetal interventions such as amniodrainage, cyst aspiration, vesicoamniotic shunt, laser vessel ablation and alcohol sclerosis can be performed to aid delivery and prevent obstructive uropathy developing in the fetus with reported long-term survival of 50%. Open fetal surgery and resection is usually unrealistic in most cases.

PREOPERATIVE PREPARATION AND ANESTHESIA

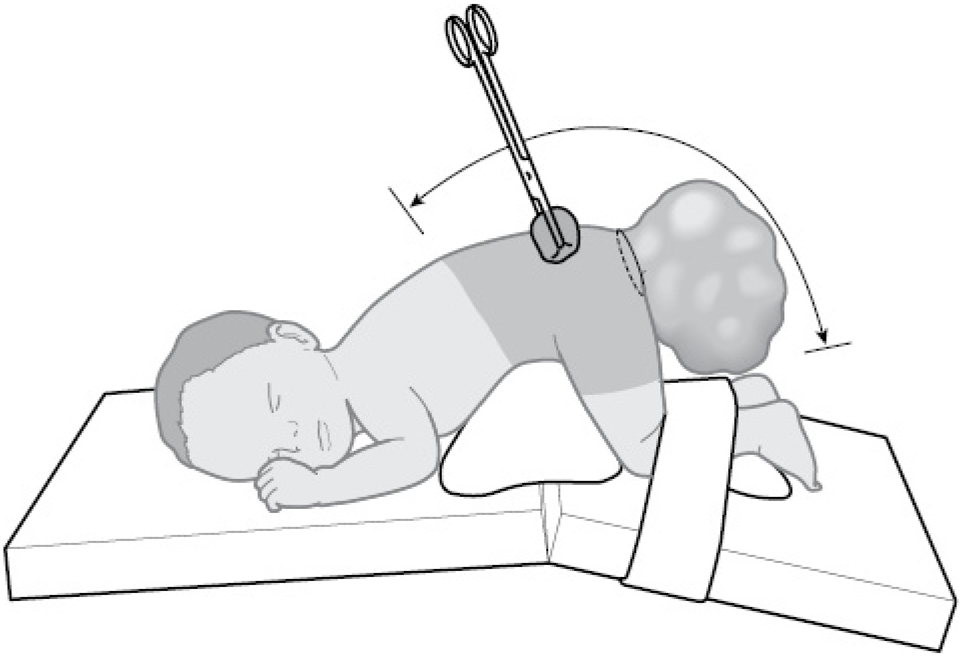
Adequate IV access and blood products should be secured before starting the operation, especially with large tumors, where there may be brisk intraoperative blood loss. In infants with SCTs with large feeding vessels or highly vascularized tumors, preoperative embolization of the feeding vessels or laparoscopic interruption of the median sacral artery can be used prior to resection to help prevent/reduce blood loss.

Although general anesthetic is mandatory, high-output cardiac failure secondary to arteriovenous channels in the tumor may limit the use of inhalation agents, which have known cardiodepressant effects.

OPERATION

Position of patient and preparation

The major component of the tumor is extrapelvic in most cases and therefore the procedure is performed with the infant in the prone “jack-knife” or “skydiver” position (pelvic and shoulder support with rolled towels) (Figure 62.3).

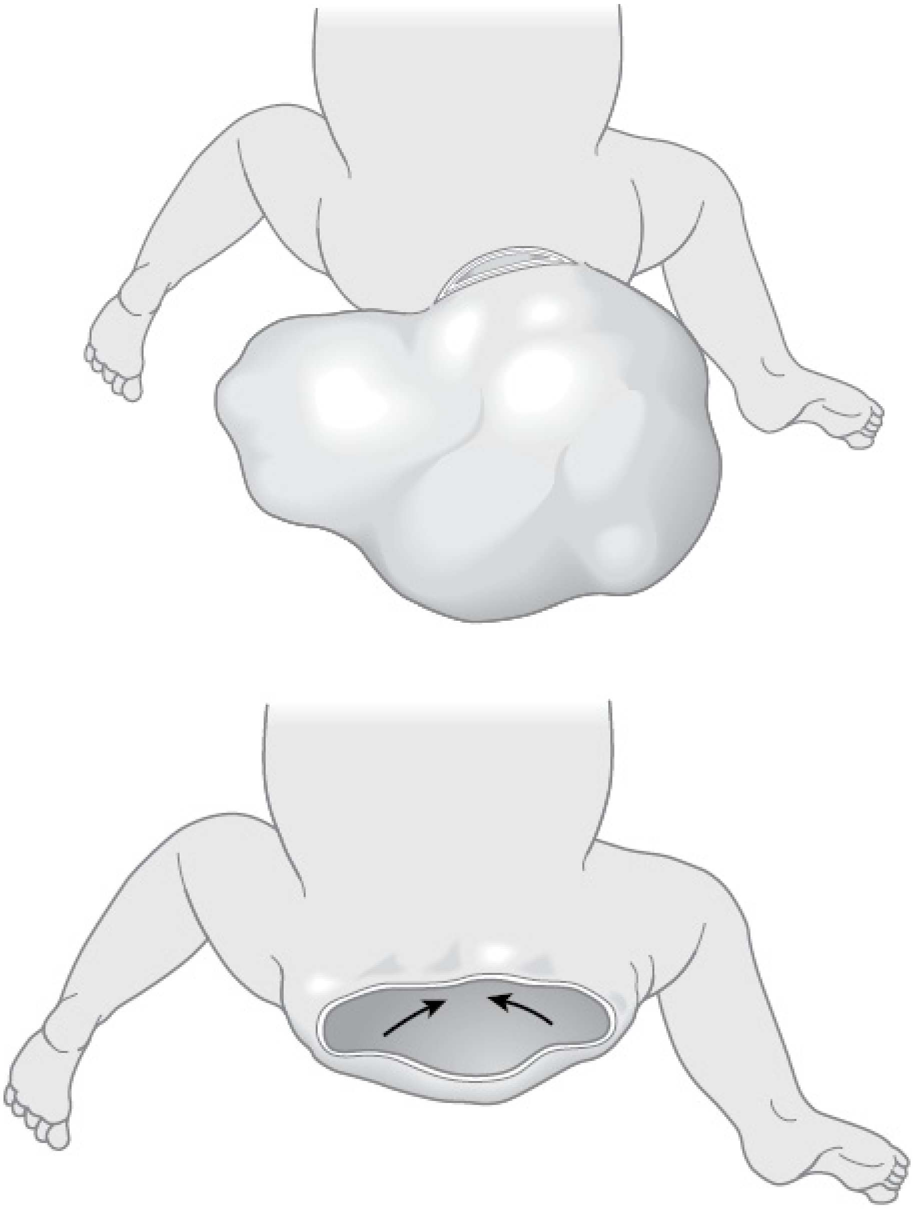


62.3

Insertion of a Hegar dilator into the rectum or packing with ribbon gauze facilitates its identification throughout the procedure.

Incision

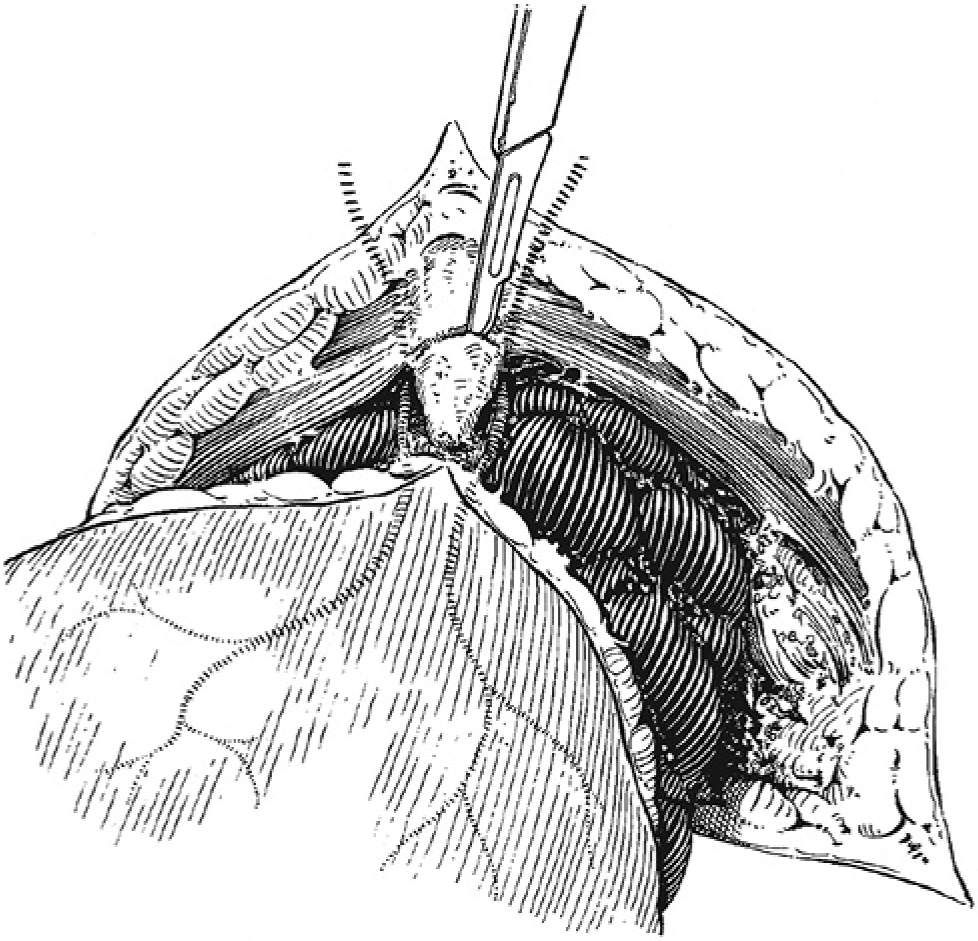
The traditional "chevron" incision is modified as this may leave protuberant "dog ears" and extend across and below the infragluteal creases down onto the posterior thighs, causing undesirable buttock deformity. Usually, the incision is simply circumferential and then at the end can be reconstructed to leave a more cosmetic appearance (**Figure 62.4a and b**).



62.4a,b

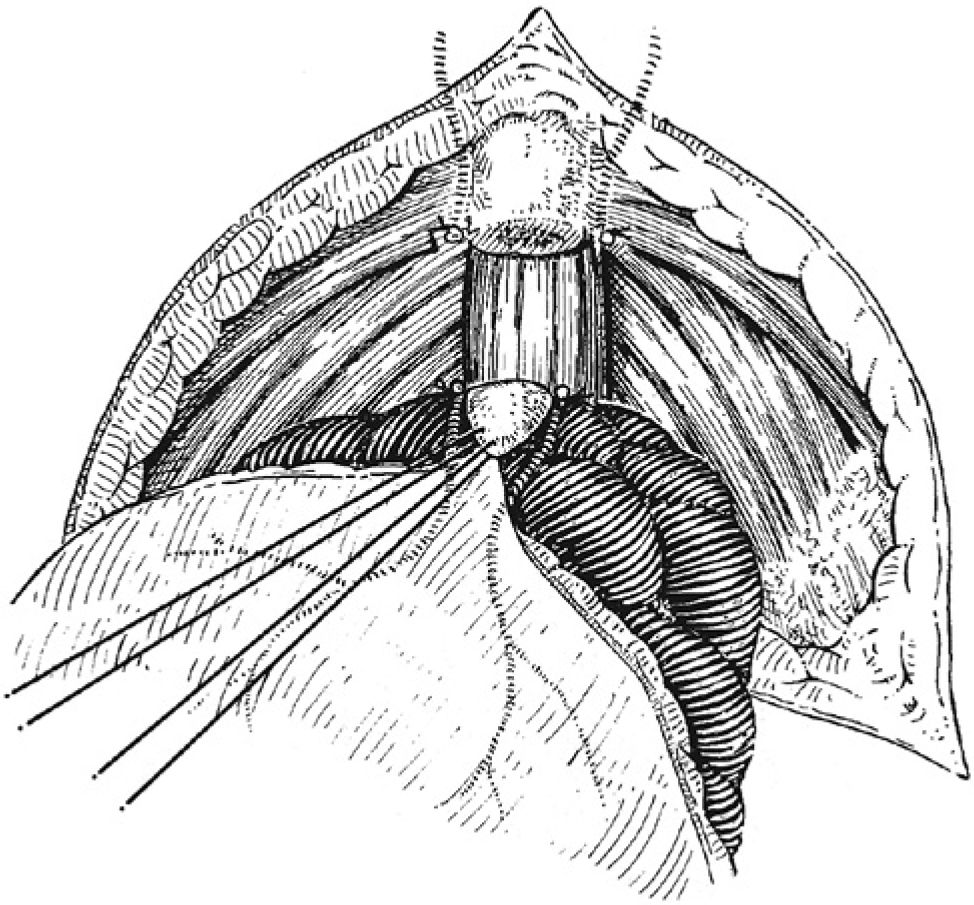
Tumor excision and coccygectomy

Skin flaps are raised to expose the stretched out gluteus muscles. The plane of the dissection is directly on the capsule of the tumor (**Figure 62.5**). Sometimes it is easier to identify the sacrum and coccyx early in the dissection; in others this can be left until later. Either way, the coccyx is transected at the sacrococcygeal joint by cautery or knife, taking the coccyx en bloc with the tumor.



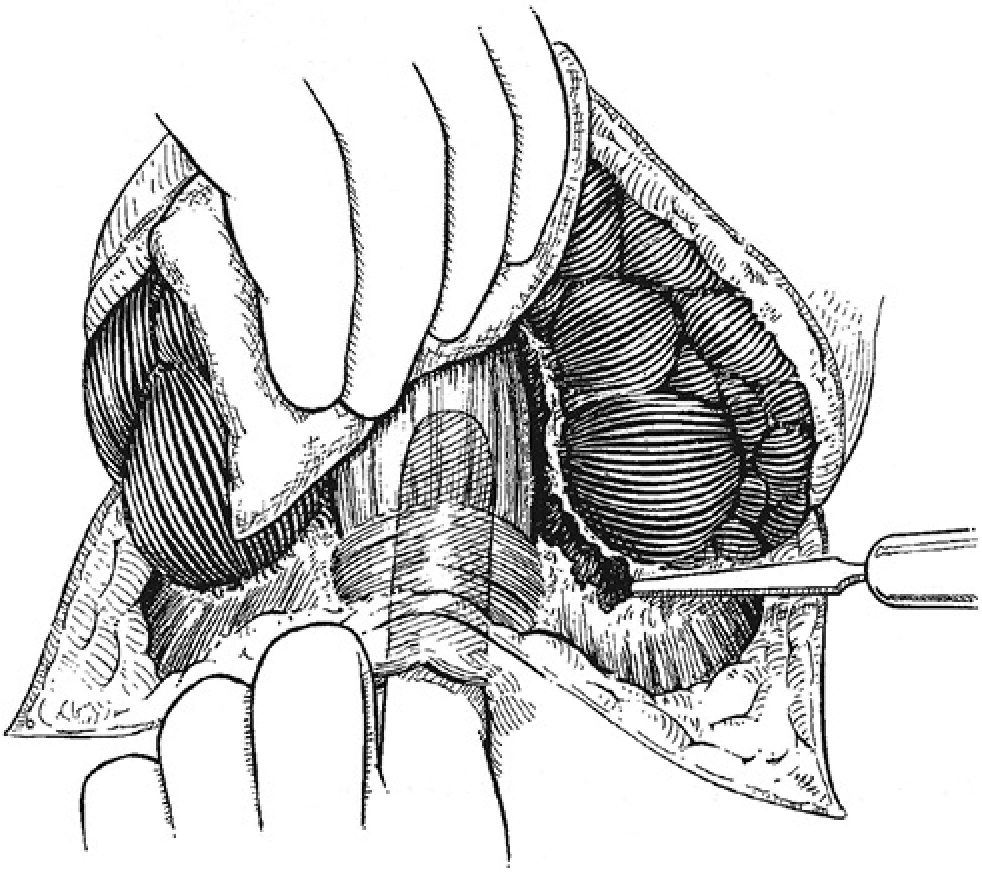
62.5

Median and lateral sacral vessels should be identified at this junction and can be coagulated or ligated with the tumor/coccyx. An initial transabdominal approach may be required to mobilize those tumors with extensive intrapelvic or abdominal extension. Using gentle traction, the coccyx and tumor are retracted and dissected free from the gluteus muscles and surrounding tissues, to the floor of the pelvis (**Figure 62.6**). Pelvic floor muscles are preserved and the proximal rectum identified. The tumor must be dissected and freed completely from the rectal wall and anorectal sphincter complex.



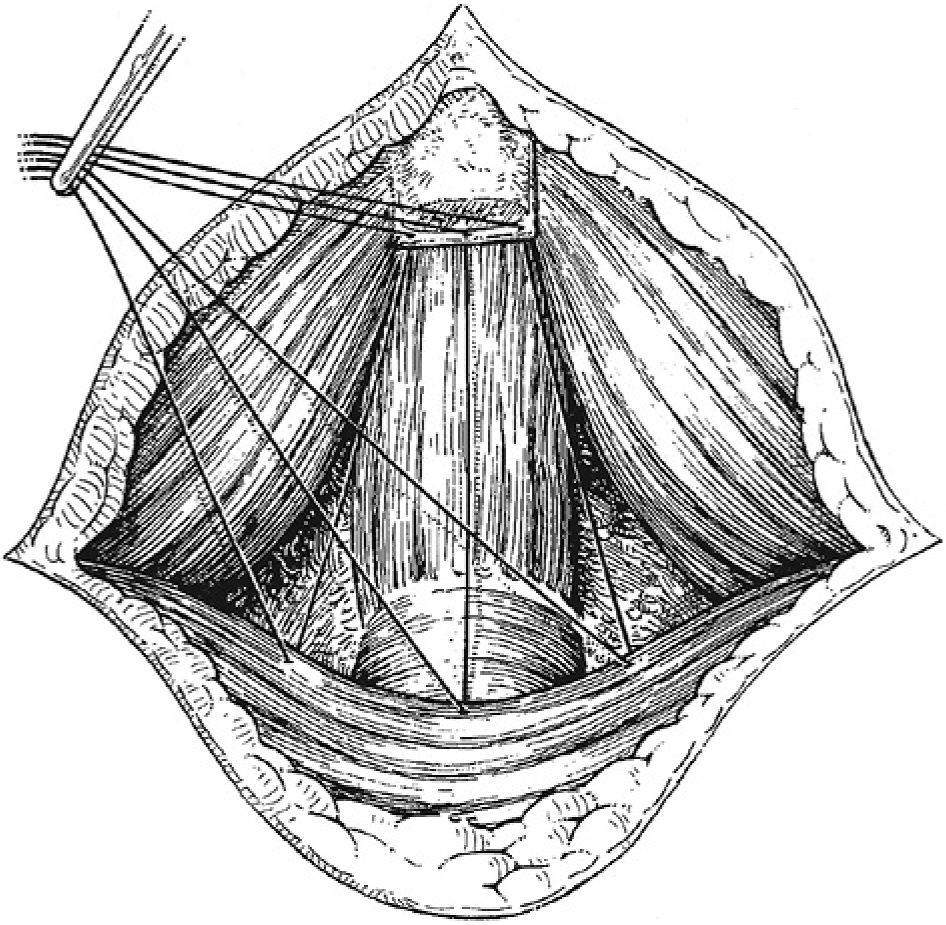
62.6

It is essential to leave the external sphincter skin and anorectal complex undamaged during mobilization of the tumor (**Figure 62.7**), though it is only rarely that it completely surrounds the rectum.



62.7

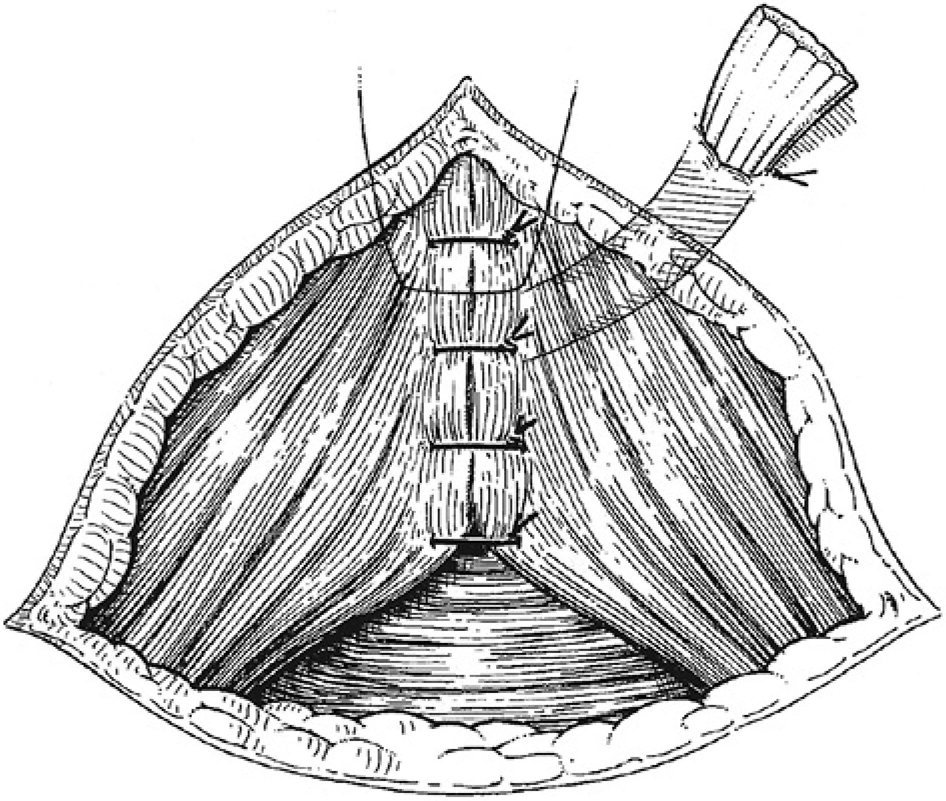
The pelvic floor is reconstructed by suturing the superior and posterior portions of the levator muscles to the presacral fascia, behind the rectum (**Figure 62.8**). This allows the anus to assume a near-normal configuration and therefore the best possibility of achieving good fecal continence.



62.8

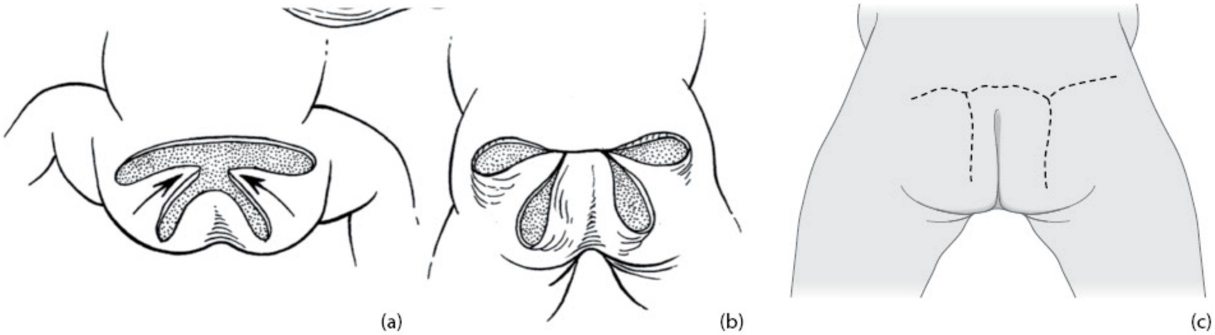
Closure

The gluteal muscles are opposed in the midline using interrupted absorbable sutures (**Figure 62.9**). Suction drain(s) may be left in place in the perirectal space prior to this closure, and brought out through a separate stab incision.



62.9

After resection of a large SCT there will be redundant skin. This will need to be resected. The ideal configuration should leave a decent buttock volume, symmetrical scars, and a median gluteal cleft. Scars that extend below the infragluteal clefts should be avoided. **Figure 62.10a–c** illustrate a reasonable method with the final appearance resembling the Greek letter π.



62.10a–c

Modifications for extensive or intra-abdominal tumors

In type III tumors where there is significant intra-abdominal involvement, the pelvic excision must be preceded by a laparotomy. In these cases, the operation commences with the patient in a supine position, with a rolled gauze pad under the buttocks, thus raising the pelvic floor. A lower midline or transverse lower abdominal incision approach is used, depending on the extent of the intra-abdominal component of the tumor. The tumor is dissected free from the lower abdominal and pelvic viscera, and the sacral vessels can be controlled through this approach. The abdominal wall is then closed, and the patient is turned prone to complete the operation as described above.

Laparoscopic approach

Laparoscopy can be used for the ligation of the median sacral vessels the resection or mobilization of intra-abdominal or pelvic portions of the SCT. Ports are placed at the umbilicus (for camera), and lateral to the umbilicus for instruments. This is combined with the external resection of the tumor, resection of the coccyx, and litigation of the lateral sacral vessels.

POSTOPERATIVE CARE

Immediate postoperative period

The child is kept in a prone position for 3 days to prevent soiling the wound with urine or feces. Oral feeding may be commenced when nasogastric aspirates are minimal, and the nasogastric tube may be removed thereafter. A pelvic neuropraxia may occur in the early postoperative period and may cause a poorly contracting, neurogenic bladder. Postoperative US should confirm complete bladder emptying prior to discharge. Although this is generally temporary, the infant will require intermittent catheterization until it resolves. The drain is removed after 48 h following drainage is minimal. In small type I tumors, the urinary catheter may be removed after

24 h, but in larger pelvic tumors abnormal bladder function and wound healing may require longer urinary catheter drainage.

Chemotherapy

Malignant lesions respond poorly with surgery alone, with only a 10% salvage rate. Adjuvant chemotherapy, particularly platinum-containing regimens such as cisplatin, bleomycin, vinblastine, and/or VP-16 (etoposide), addressing the specific malignant element, has improved survival with such lesions. This regimen may shrink the tumor, making it amenable to secondary resection. All resected tumors should be examined for malignant elements, which may require subsequent chemotherapy.

Follow-up

Beyond the immediate postoperative care period, careful monitoring is required as malignant germ-cell tumors can recur either from missed malignant elements in the original tumor or from malignant conversion of benign residual tissue. Follow-up, including rectal examinations and abdominal US, should be performed at monthly intervals for the first year, at 3-month intervals for at least 3 years (as most recurrences occur within the first 3 years), and then annually. Monitoring of serum AFP and β-hCG levels may also help detect the presence of malignant recurrence. Recurrent tumors should be excised, with preoperative chemotherapy for more extensive disease.

COMPLICATIONS AND OUTCOME

The prognosis for patients with SCT is dependent on the following:

- *Antenatal/obstetric factors:* Hydrops or placentomegaly is associated with almost 100% mortality, while dystocia or tumor rupture during delivery may be associated with exsanguinating hemorrhage.

- *Age at diagnosis:* The risk of malignant transformation in a benign lesion increases with age.
- *Tumor histology and stage:* Up to 95% of benign tumors can be cured with excision surgery alone. Prognoses of malignant tumors are dependent on tumor type, stage, and location. Survival with malignant tumors can be achieved in up to 90% of cases using a combination of surgery and adjuvant chemotherapy, although the risk of late recurrences or second malignancies persists. Following surgical excision, mature teratomas are associated with an 11% risk of recurrence within 5 years.
- *Surgical factors:* Recurrence is more likely with incompletely excised lesions or in the absence of coccygectomy. Failure to remove the coccyx is associated with a 30% incidence of recurrence, with more than 50% of cases being malignant.

While the overall outlook for resected SCT is good, long-term complications may be seen including mild bowel dysfunction (incontinence or more likely constipation), a degree of urinary incontinence, or neuropathic bladders. Gait or locomotor problems are, perhaps surprisingly, uncommon. Long-term surveillance is recommended.

FURTHER READING

Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey, 1973. *J Pediatr Surg* 1974; 9: 389–98.

Draper H, Chitayat D, Ein SH, Langer JC. Long-term functional results following resection of neonatal sacrococcygeal teratoma. *Pediatr Surg Int* 2009; 25: 243–6.

Fishman SJ, Jennings RW, Johnson SM, Kim HB. Contouring buttock reconstruction after sacrococcygeal teratoma resection. *J Pediatr Surg* 2004; 39: 439–41.

Flake AW. Fetal sacrococcygeal teratoma. *Semin Pediatr Surg* 1993; 2: 113–20.

Hambraeus M, Al-Mashhadi A, Wester T, et al. Functional outcome and health-related quality of life in patients with sacrococcygeal teratoma – a Swedish multicenter study. *J Pediatr Surg* 2019; 54(8): 1638–43.

Le LD, Alam S, Lim FY et al. Prenatal and postnatal urological complications of sacrococcygeal teratomas. *J Pediatr Surg* 2011; 46: 1186–90.

Makin EC, Hyett J, Ade-Ajayi N, et al. Outcome of antenatally diagnosed sacrococcygeal teratomas: Single-center experience (1993–2004). *J Pediatr Surg* 2006; 41(2): 388–93.

Sebire NJ, Fowler D, Ramsay AD. Sacrococcygeal tumors in infancy and childhood; A retrospective histopathological review of 85 cases. *Fetal Pediatr Pathol* 2004; 23: 295–303.

PART VI

Endocrine

63

Thyroidectomy

STEVEN W. BRUCH

HISTORY

Although the first recorded thyroidectomy occurred in 952 AD, it was not until the 1870s that thyroidectomies were more routinely performed. Theodor Billroth and his pupil Theodor Kocher led the development of thyroid surgery, bringing the mortality rate down from over 40% prior to 1850 to Kocher's 0.5% rate over his career of nearly 5000 thyroidectomies. William Halstead, after working with both Billroth and Kocher, became the most prominent early American authority on thyroid surgery. Halstead noted that Billroth's patients almost never developed myxedema but often suffered tetany, whereas Kocher had the opposite experience. Halstead theorized that "the explanation probably lies in the operative methods of the two illustrious surgeons. Kocher, neat and precise, operating in a relatively bloodless manner, scrupulously removed the entire thyroid gland, doing little damage outside the capsule. Billroth, operating more rapidly, and as I recall his manner, with less regard for tissue and less concern for hemorrhage, might easily have removed the parathyroids or at least interfered with their blood supply, and have left remnants of thyroid."

PRINCIPLES AND JUSTIFICATION

Children require a thyroidectomy for three main etiologies: Grave's disease, prophylactic thyroidectomies for multiple endocrine neoplasia (MEN) type

2, and thyroid nodules with a risk of cancer.

Grave's disease causes hyperthyroidism when autoantibodies stimulate the TSH receptor on thyroid follicular cells resulting in increased vascularity, follicular hyperplasia, and increased synthesis and secretion of thyroid hormone. Grave's disease is the most common cause of hyperthyroidism in children, but it occurs rarely, in about 1 in 100 000 children. Medical treatment with methimazole and beta blockers provides initial treatment. If ineffective, radioactive iodine ablation or, preferably in children, a total thyroidectomy is required.

Patients with MEN 2 syndromes require a prophylactic thyroidectomy since they all develop medullary thyroid cancer. MEN 2A and 2B are both a result of a mutation of the *RET* gene. MEN 2A patients develop hyperparathyroidism and pheochromocytomas in addition to medullary thyroid cancer, while MEN 2B patients develop pheochromocytomas, medullary thyroid cancer, and ganglioneuromatosis. The *RET* mutation determines the degree of aggressiveness of the medullary thyroid cancer and thus the optimal age for thyroidectomy. The patients are divided into three groups depending on their mutation: Highest risk (codon 918), high risk (codons 634 and 883), and moderate risk (all other codons). Patients in the highest risk category are the MEN 2B patients who require a prophylactic total thyroidectomy before the age of 1 year. High- and moderate-risk patients are MEN 2A patients. The high-risk patients require their prophylactic thyroidectomy at or before age 5 depending on calcitonin levels. Moderate-risk patients have their calcitonin levels followed, and undergo a total thyroidectomy when the calcitonin begins to rise.

Thyroid nodules are less common in children (2%) than adults (25%), but the nodule's risk of malignancy is greater than that of a nodule discovered in an adult (22–26% compared to 5–10%). The incidence of thyroid cancer is tenfold greater in adolescents than younger children and there is a 5 : 1 female to male preponderance. Fine-needle aspiration and the Bethesda System for Reporting Thyroid Cytopathology determine the risk of cancer in thyroid nodules and thus helps determine the surgical treatment. Table 63.1 shows the Bethesda classifications, their risk of cancer in children, and the recommended surgical procedure.

Table 63.1 Bethesda classification, cancer risk, and recommended surgical intervention in children

Bethesda classification	Cancer risk in children (%)	Surgical intervention
Non-diagnostic	NA	Repeat fine-needle aspiration
Benign	7	Observe; if nodule >4 cm, lobectomy
Atypia or follicular lesion of uncertain significance	28	Lobectomy
Follicular or Hurthle cell neoplasm	58	Lobectomy
Suggestive of malignancy	85–100	Lobectomy or total thyroidectomy
Malignant	100	Total thyroidectomy

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

The preparation of patients with Grave's disease, MEN 2, and thyroid nodules varies. Grave's patients are often on methimazole and may require a beta blocker to control their hyperthyroidism. Occasionally, levothyroxine is added if the methimazole produces a hypothyroid state. In conjunction with a pediatric endocrinologist, all attempts are made to make the patient euthyroid prior to the procedure. To prevent thyroid storm and to make the gland less vascular, patients begin a beta blocker and Lugol's iodine 10 days to 2 weeks prior to the operation. The Lugol's iodine is stopped and the beta blocker is weaned after the operation.

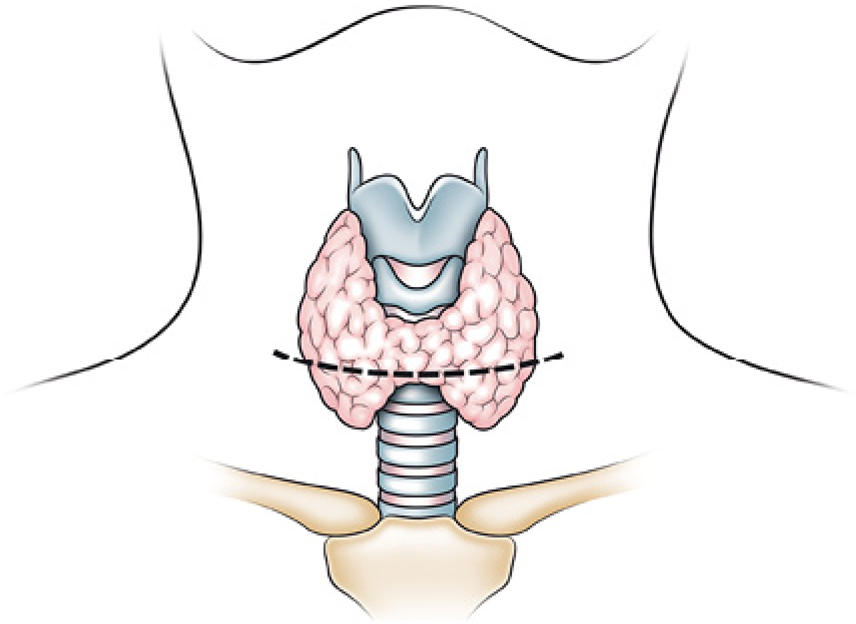
MEN 2 patients undergo ultrasonography of the neck, and are followed with serum calcitonin levels in preparation for their prophylactic thyroidectomy. An elevation in calcitonin or an abnormal ultrasound (US) may prompt the operation to be performed prior to its originally planned date.

Patients with thyroid nodules present euthryoid in the majority of cases. The occasional hyperfunctioning nodule that presents with a low TSH, and is "hot" on radioactive iodine scan requires a lobectomy according to the American Thyroid Association (ATA) guidelines. These patients should be made euthyroid prior to lobectomy with methimazole and possibly a beta blocker. Prior to surgery for suspected cancer, US to look at lateral neck nodes and fine-needle aspiration biopsy with a thyroglobulin wash of abnormal neck nodes helps determine the extent of surgery. Positive lateral neck nodes require the addition of a modified radical neck dissection to the total thyroidectomy and central neck lymph node dissection. The US can also be useful to look at the carotid–subclavian junction on the right. If the junction is abnormal, there is a higher likelihood of discovering a non-recurrent recurrent laryngeal nerve.

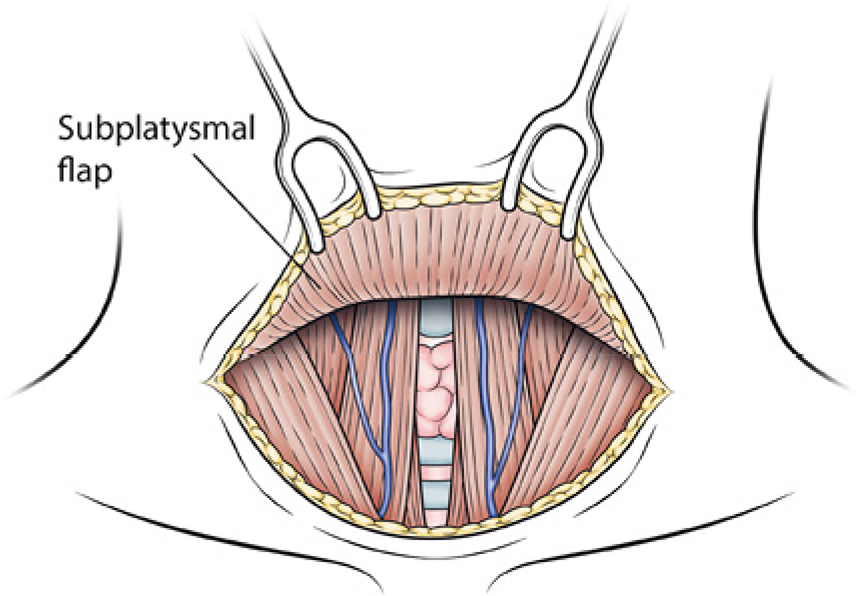
OPERATION

The patient is anesthetized and a NIM® endotracheal tube is placed under direct vision if recurrent laryngeal nerve monitoring is utilized. The patient is positioned with a bump under the scapula and the head extended and padded. A modified Beech-chair position with the upper half of the operating table elevated at a 45-degree angle decreases venous pressure.

A curvilinear incision in the lower neck hidden in the skin lines over the isthmus of the thyroid gland is brought through the skin and platysma muscle (**Figure 63.1**). Subplatysmal flaps are created superiorly to the thyroid cartilage and inferiorly to the sternal notch (**Figure 63.2**). The strap muscles are separated in the midline and then the sternohyoid and sternothyroid muscles are separated laterally. If nerve monitoring is used, the vagus nerve should be isolated and stimulated in the carotid sheath to ensure the monitoring system is in working order.

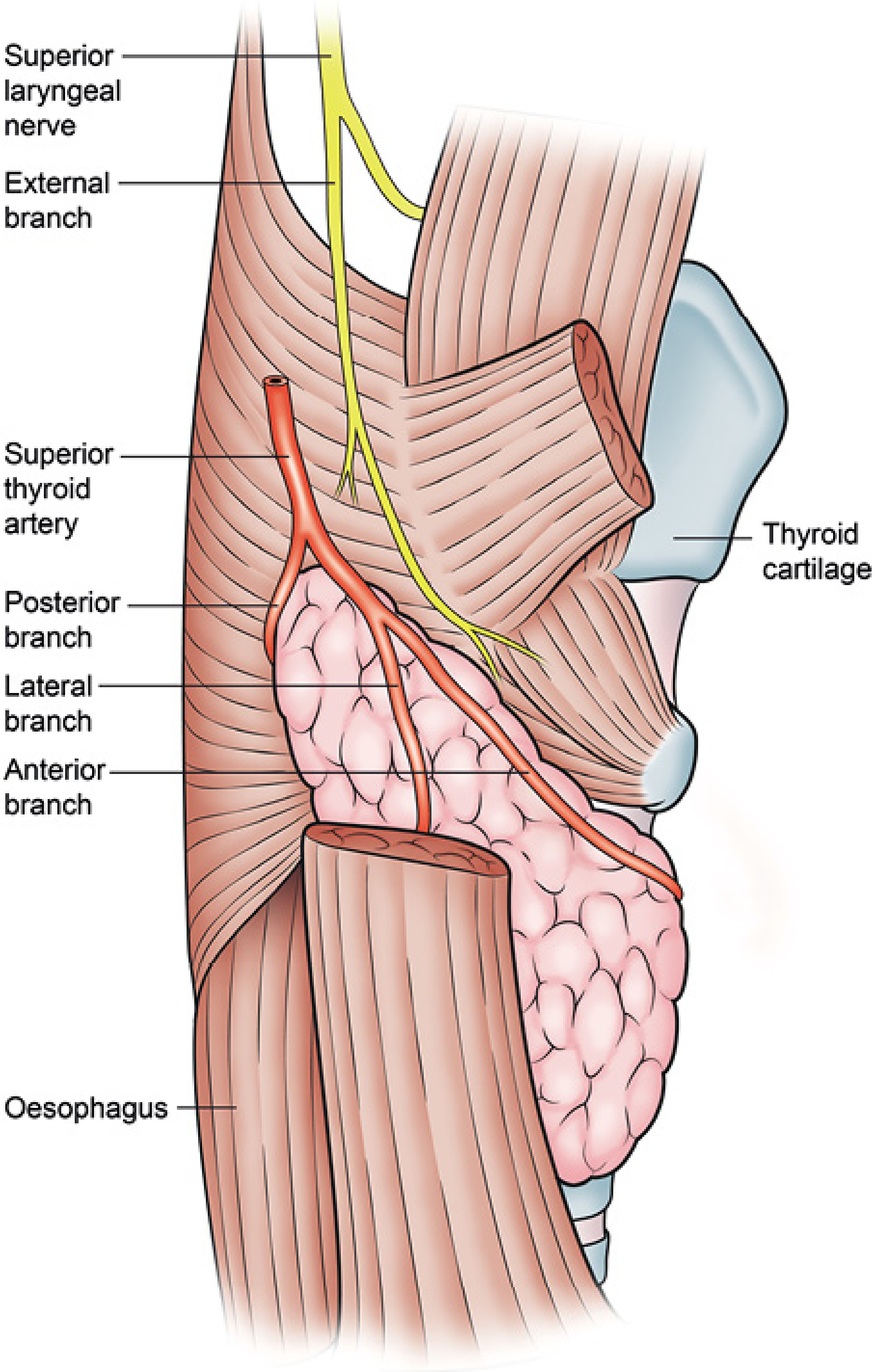


63.1

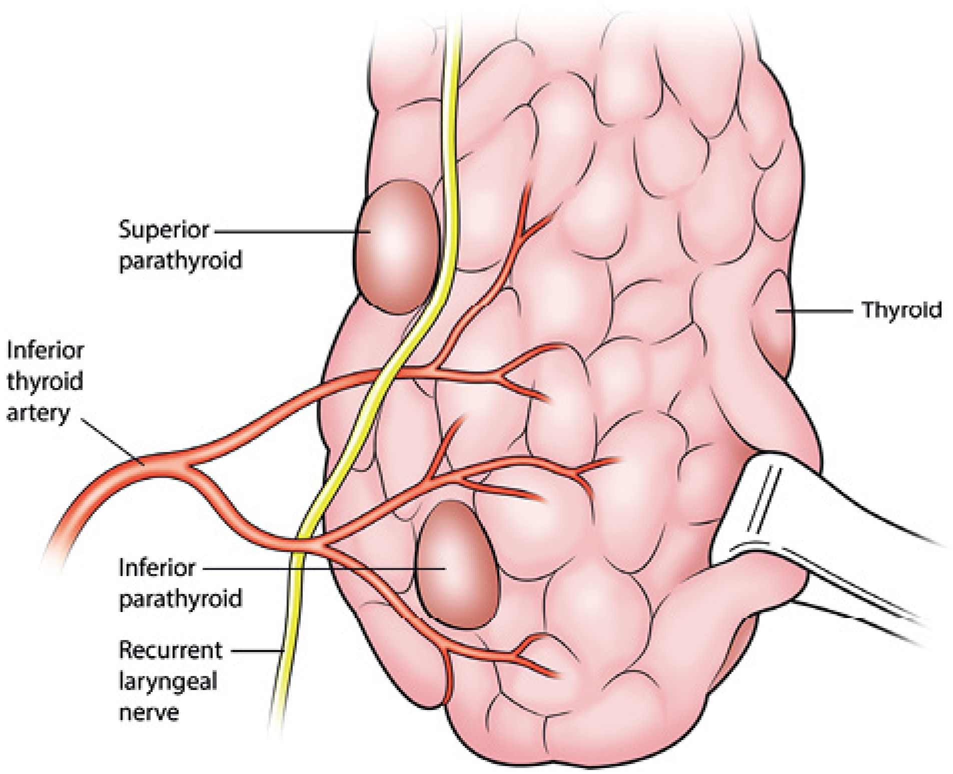


63.2

The superior pole vessels are then divided individually at the level of the thyroid capsule (**Figure 63.3**). This avoids injury to the superior laryngeal nerve. The gland is then rotated lateral to medial and the middle thyroid vein is divided again at the level of the thyroid capsule. Staying on the capsule of the gland, the remaining vasculature is controlled while identifying the superior and inferior parathyroid glands and the recurrent laryngeal nerves (**Figure 63.4**).


Superior laryngeal nerve
External branch
Superior thyroid artery
Posterior branch
Lateral branch
Anterior branch
Oesophagus
Thyroid cartilage

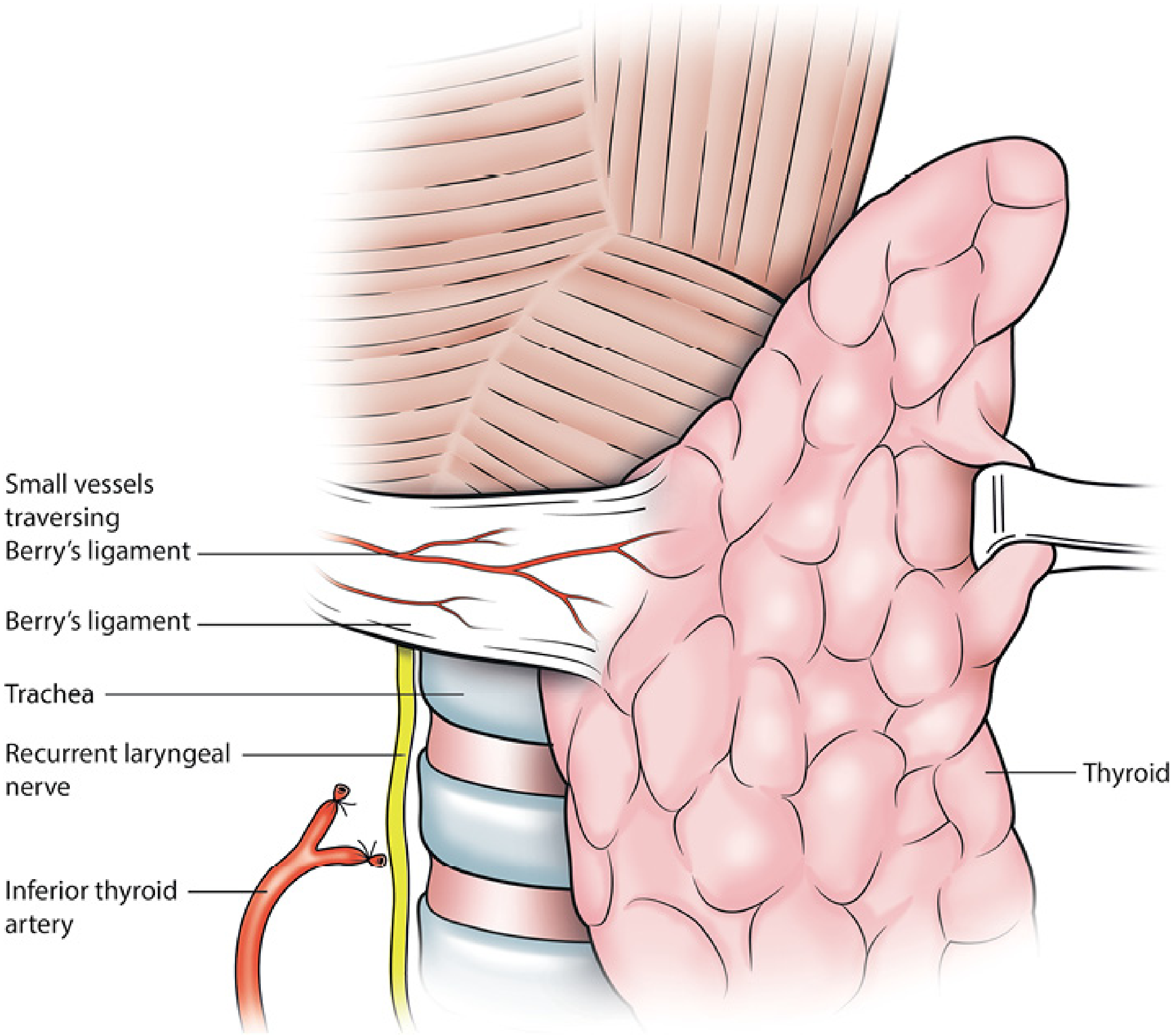
63.3



63.4

The superior parathyroid gland is usually found lateral to the recurrent laryngeal nerve at the level of the cricoid cartilage. The inferior parathyroid glands most often are located anterior to the recurrent laryngeal nerve and caudal to the intersection of the recurrent laryngeal nerve and the inferior thyroid artery. The recurrent laryngeal nerve has a slightly different course on the right side compared to the left. On both sides the recurrent laryngeal nerves enter the larynx posteromedially in the cricothyroid muscle just superior to the cricoid cartilage. The recurrent laryngeal nerve wraps around the innominate artery on the right and enters the larynx from a more lateral to medial direction than does the left recurrent laryngeal nerve, which wraps around the aortic arch deeper in the chest and runs more parallel in the tracheoesophageal groove.

The thyroid is then dissected from the anterior aspect of the trachea. This dissection is most difficult at Berry's ligament, where the recurrent laryngeal nerve usually is entering the larynx (**Figure 63.5**). There are often small vessels close to the nerve in this area that, if they are not initially controlled, should be handled with pressure and hemostatic agents to avoid nerve injury. With a lobectomy, the isthmus is dissected over to the opposite lobe and removed entirely; a total thyroidectomy entails the same maneuvers on the opposite side. Prior to leaving the initial operative side, the vagus nerve should again be stimulated to ensure an intact recurrent laryngeal nerve if the nerve monitoring system is being used. Following completion of the thyroid resection, hemostasis is ensured. The strap muscles are brought together in the midline with interrupted sutures, leaving a generous opening inferiorly for blood to escape if a postoperative hematoma were to develop. The platysma muscle is brought together, followed by the subcuticular layer and the skin.


Small vessels traversing Berry's ligament
Berry's ligament
Trachea
Recurrent laryngeal nerve
Inferior thyroid artery
Thyroid

63.5

POSTOPERATIVE CARE

Postoperative care centers around two complications and their management: Neck hematoma, and hypocalcemia. A neck hematoma presents with neck swelling, difficulty breathing, and a feeling of impending doom by the patient. If this occurs, the neck incision should be opened at the bedside to relieve the hematoma while an airway is being secured. It is advisable to send an instrument tray with the patient from the operating room to the ward to avoid a delay in opening the neck. Once the pressure of the hematoma is relieved, the patient should be brought to the operating room to wash out the wound and identify the inciting bleeder.

Hypocalcemia presents with symptoms of perioral and fingertip numbness and tingling, along with a positive Chvostek's and/or Trousseau's sign. This rarely occurs after a thyroid lobectomy, but it is not uncommon after a total thyroidectomy, and occurs more often following a central neck dissection. This results from inadvertent removal or disruption of the blood supply to the parathyroid glands during the dissection. Potential hypocalcemia can be dealt with in one of two ways: By routine supplementation of all total thyroidectomies with oral calcium, or checking a serum parathyroid hormone (PTH) level 4 hours after the operation is completed. If the serum PTH level is normal, no calcium supplementation is required, and, if it is low, the supplementation is started. On occasion, IV calcium with calcitriol is required to control hypocalcemia in the hospital prior to outpatient supplementation.

The hospital stay is rarely longer than 24 hours.

COMPLICATIONS

Three major complications can occur following thyroid surgery in children: Bleeding, recurrent laryngeal nerve injury, and parathyroid insufficiency resulting in hypocalcemia. Postoperative neck hematoma occurs in up to 1% of cases and is avoided by meticulous hemostasis.

Similarly, recurrent laryngeal nerve injury occurs infrequently in 1–2% of cases. Again, meticulous dissection, with or without the aid of recurrent laryngeal nerve monitoring, helps avoid this complication. If there is a suspected recurrent laryngeal nerve injury during the first side of a total thyroidectomy, one should stop at that point, evaluate the vocal cords postoperatively, and consider removing the other side at a later date depending on pathology results and the status of the vocal cords with time. If a nerve transection is recognized during the dissection, an attempt to repair the nerve with fine suture and the aid of an operating microscope should be made.

Hypocalcemia rarely occurs with a thyroid lobectomy. However, transient hypocalcemia occurs in 14–34% of children undergoing a total thyroidectomy. The hypocalcemia resolves in the majority of these children within a month after the operation. However, 1–4% of children will remain hypocalcemic at a year and require long-term calcium and calcitriol replacement. In 20–24% of total thyroidectomies in children, a parathyroid gland is removed inadvertently. If this is recognized, the gland should be divided into small slivers and re-implanted in the sternocleidomastoid muscle or the forearm. Risk factors for hypocalcemia include total thyroidectomy with lymph node dissection, younger age, and patients with hyperthyroidism.

OUTCOME

Although thyroid nodules occur uncommonly in children, there is a greater risk of malignancy in nodules found in children compared to adults. In children 22–26% of nodules harbor malignancy compared to only 5–10% in adults. Children more often have disease extension outside the thyroid, regional lymph node involvement, and pulmonary metastasis compared to adults, even after controlling for tumor size and pathology. But, despite presenting with more extensive disease, children die from their thyroid cancer much less often (less than 2% long-term disease-specific mortality) compared to adults.

FURTHER READING

DuBose J, Barnett R, Ragsdale T. Honest and sensible surgeons: The history of thyroid surgery. *Curr Surg* 2004; 61: 213–19.

Francis GL, Waguespack SG, Bauer AJ, et al. Management guidelines for children with thyroid nodules and differentiated thyroid cancer. The American Thyroid Association Guidelines Task Force on Pediatric Thyroid Cancer. *Thyroid* 2015; 25: 716–59.

Srinivasan S, Misra M. Hyperthyroidism in children. *Pediatr Rev* 2015; 36: 239–47.

Wells SA Jr, Asa SL, Dralle H, et al. Revised American Thyroid Association guidelines for the management of medullary thyroid carcinoma. The American Thyroid Association Guidelines Task Force on Medullary Thyroid Carcinoma. *Thyroid* 2015; 25: 567–610.

64

Pancreatectomy for hyperinsulinism

N. SCOTT ADZICK AND PABLO LAJE

INTRODUCTION

Congenital hyperinsulinism (HI) is a heterogeneous genetic disorder of the insulin metabolism that results in severe persistent neonatal hypoglycemia. There are two major histological forms: Focal HI, in which only a discrete area of the pancreas is affected, and diffuse HI, in which the entire organ is diseased. Partial pancreatectomy is indicated as a curative procedure in all cases of focal HI. On the other hand, near-total pancreatectomy is indicated in some genetic variants of diffuse HI, with the goal of decreasing the severity of the disease. While the operation is highly effective in achieving this goal, it is generally rendered as a palliative procedure since only about 30% of patients who undergo a near-total pancreatectomy achieve a definitive cure and need no postoperative glucose management. Certain genetic variants of diffuse HI do not need surgery and are managed medically. If untreated, all forms of HI result in irreversible brain damage and neurodevelopmental delay. There are about 80 to 120 cases of HI per year in the United States. Due to their scarcity, patients with HI should only be managed in centers with an expert multidisciplinary team.

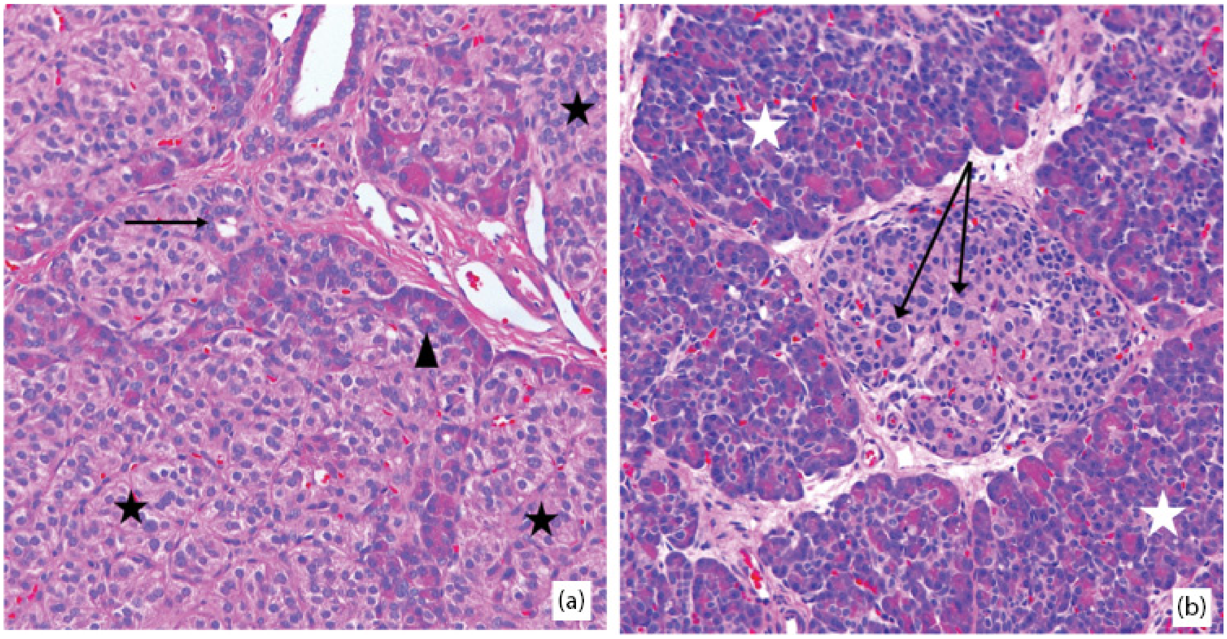
HISTORY

The first successful pancreatectomy in a child with HI was performed in 1934 by Evarts Ambrose Graham, an American surgeon working in St

Louis, Missouri, USA. Searching for what was thought to be an adenoma, a subtotal pancreatectomy was done and the patient's hypoglycemia resolved. The operation was done 20 years before the first description of the disease by Irvine McQuarrie in 1954, and was initially termed "syndrome of idiopathic hypoglycemia of infants." In the 1970s the disease was named "nesidioblastosis," a term abandoned in the 1990s after advances in molecular diagnosis showed that it results from a variety of genetic derangements that alter the regulatory mechanisms of insulin secretion and glucose homeostasis. The term "congenital hyperinsulinism" is the current term for the family of diseases characterized as a heterogeneous genetic disorder of insulin metabolism that results in severe persistent neonatal hypoglycemia, often associated with neurological complications.

PRINCIPLES AND JUSTIFICATION

There are two major histological forms of HI, focal (**Figure 64.1a**) and diffuse (**Figure 64.1b**), which have a different genetic background and management strategy.



64.1a,b (a) Focal lesion. There is hyperplasia of beta cells (black stars), with exocrine (black arrowhead) and ductal (black arrow) components within the lesion. (b) Diffuse HI. Endocrine cells show nucleomegaly (black arrows) within an otherwise normal islet of Langerhans.

Focal HI (~40% of all patients) consists of a single focus of adenomatous islet cell hyperplasia surrounded by normal pancreatic tissue. Focal lesions vary in size from a few millimeters to greater than a centimeter, and can be on the surface or deep within the pancreas. About 50% of lesions are located in the pancreatic head, 25% in the neck/body, 15% in the tail, and 15% in other locations.

In diffuse HI (~60% of all patients), the appearance of the pancreas is normal except for one histological hallmark feature: Beta cell nucleomegaly. The abnormal beta cells are distributed homogeneously throughout the organ and the total number of beta cells within the organ is no different from those from healthy age-matched controls.

In our experience of more than 460 HI patients coming to surgery since 1999 (which represents about 60–70% of all HI patients treated at our center in Philadelphia), 55% had focal disease and 45% had diffuse disease.

There are rare cases that do not fit in either of the two categories and are called atypical. Among these are cases of focal lesions that occupy a large segment of the pancreas, cases with remarkable endocrine hyperplasia in the context of Beckwith–Wiedemann syndrome, and patients with features of diffuse HI restricted to a single area of the pancreas distributed in a mosaic pattern which we term "localized islet nuclear enlargement" (LINE). Patients with atypical forms of HI are clinically heterogeneous and require a medical and surgical treatment plan that is individually crafted according to the severity of each case.

Biological mechanism and diagnosis

The most common pathophysiologic mechanism is a loss-of-function genetic mutation of the ATP-dependent potassium channels (K-ATP channels) of the beta cell membrane, leading to constant and unregulated secretion of insulin. The K-ATP channel of the beta cell membrane is composed of the subunits coded by two genes located next to each other in the p15.4 region of the chromosome 11. Diffuse HI occurs most frequently due to homozygous mutations of these genes inherited in an autosomal recessive manner. Less frequently, diffuse HI occurs due to mutations in the genes of nine other enzymes and metabolic factors involved in the glucose metabolism. Focal HI occurs through a "two-hit" phenomenon. The first "hit" occurs when an individual inherits a mutation in the paternal allele in one of the K-ATP channels genes, and the second "hit" occurs when the maternal 11p15 region containing the normal maternal allele is lost. The loss of the maternal 11p15 region is a completely random event that has no inheritable component and is called "loss of heterozygosity." The affected beta cell not only oversecretes insulin but also develops an adenomatous hyperplastic proliferation due to an imbalance in a series of genes that regulate cell proliferation that are also contained in the 11p15 region and are subject to strong genomic imprinting.

When a baby is diagnosed with HI in the absence of a family history, the parents and the patient must undergo genetic testing to help differentiate between diffuse and focal HI, determine the need for imaging studies, determine the surgical approach, and provide prognostic information.

The diagnosis of HI is established through a series of three simple blood tests:

- Fasting and postprandial hypoglycemia with unsuppressed HI (neonatal hypoglycemia is defined as a glucose plasma concentration of <50 mg/dL (2.8 mmol/L) after the first 24 hours of life with a plasma insulin concentration of >36 pmol/L)
- Suppression of lipolysis and ketogenesis at the time of the hypoglycemia
- A positive hyperglycemic response to a dose of glucagon

These criteria must be present for a prolonged period of time and outside certain clinical circumstances such as perinatal stress and sepsis.

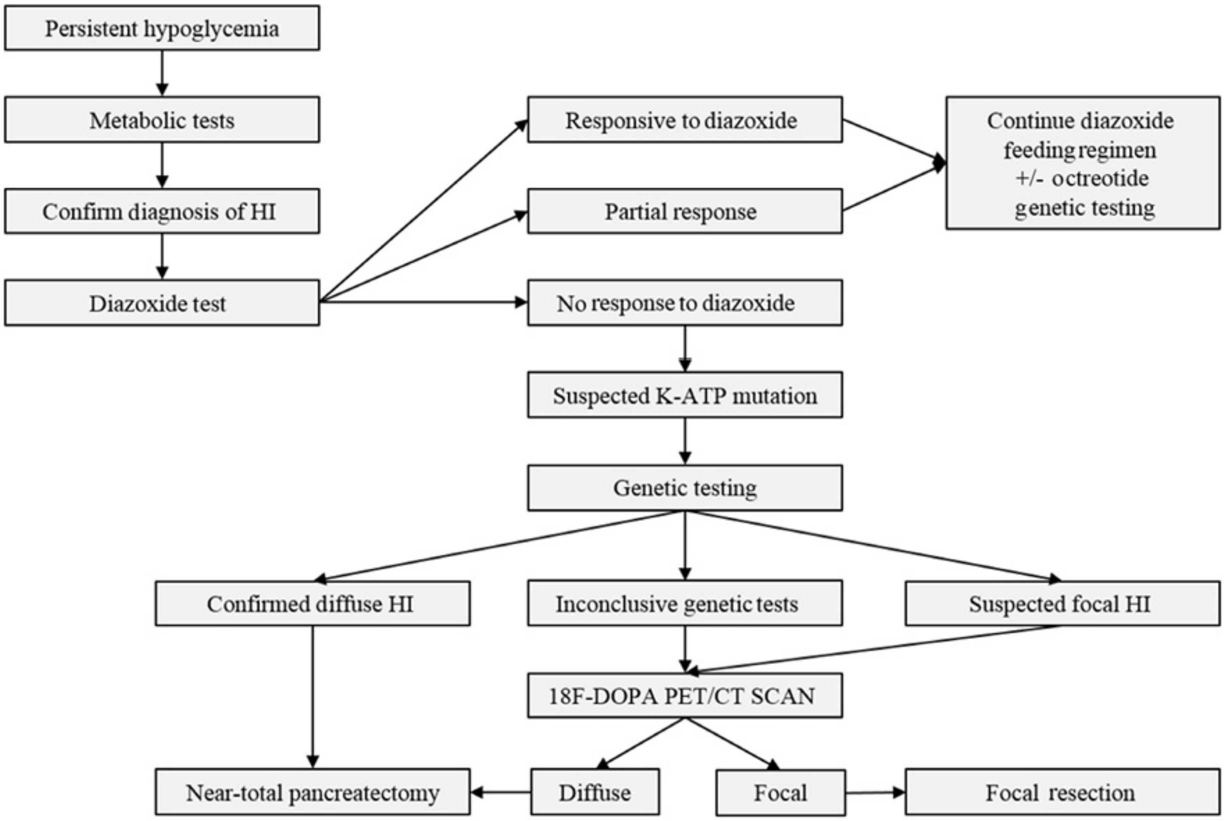
Prenatal screening of all known mutations of all HI-related genes in the general population is impractical due to the low incidence of the disease. On the other hand, prenatal diagnosis in families with affected probands is possible and justified because it allows immediate postnatal management.

THERAPEUTIC CLASSIFICATION AND MEDICAL MANAGEMENT

The mainstay drug in the treatment of HI is diazoxide, which inhibits insulin secretion by activating the K-ATP channel. In order to be effective, both subunits of the K-ATP channel must be structurally and functionally normal. From a treatment standpoint, HI is divided into two groups: Diazoxide-responsive and diazoxide-resistant. Since the most common cause of HI is a mutated *SUR1/Kir6.2* gene complex, the majority of HI patients do not respond to diazoxide. The ones that respond are those with mutations in the *GK*, *GDH*, *SCHAD*, and other HI-related genes. Patients who respond to diazoxide are managed by a regimen of frequent feedings and long-term diazoxide. Overall, about one-third of HI patients are diazoxide-responsive and two-thirds are diazoxide-resistant. Diazoxide-resistant patients generally require surgery.

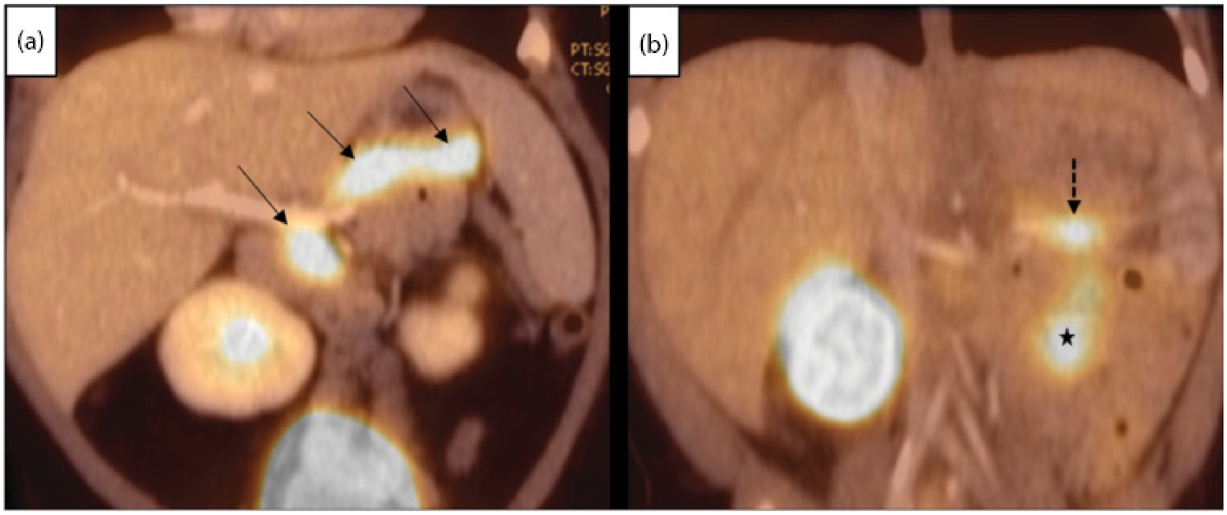
KEY PREOPERATIVE INVESTIGATIONS

The most important aspect of the preoperative planning is to determine whether the patient has diffuse HI or focal HI because the surgical strategy is radically different between the two. Genetic testing is the first step. In the ideal situation, a known disease-causing K-ATP channel mutation is found on the maternal and paternal alleles, confirming diffuse HI, or a disease-causing mutation is found in the *paternal* allele only, suggesting focal disease. When the genetic testing provides inconclusive results, the patient must undergo imaging studies to distinguish between focal and diffuse HI. Patients with genetically confirmed recessive K-ATP-related diazoxide-resistant diffuse HI do not need preoperative imaging studies. On the other hand, patients with genetic testing suggestive of focal HI must undergo imaging studies to confirm focal disease and to localize the suspected focal lesion (**Figure 64.2**).


Persistent hypoglycemia
Metabolic tests
Confirm diagnosis of HI
Diazoxide test
Responsive to diazoxide
Partial response
No response to diazoxide
Continue diazoxide
feeding regimen
+/- octreotide
genetic testing
Suspected K-ATP mutation
Genetic testing
Confirmed diffuse HI
Inconclusive genetic tests
Suspected focal HI
18F-DOPA PET/CT SCAN
Near-total pancreatectomy
Diffuse
Focal
Focal resection

64.2 Management algorithm for patients with HI.

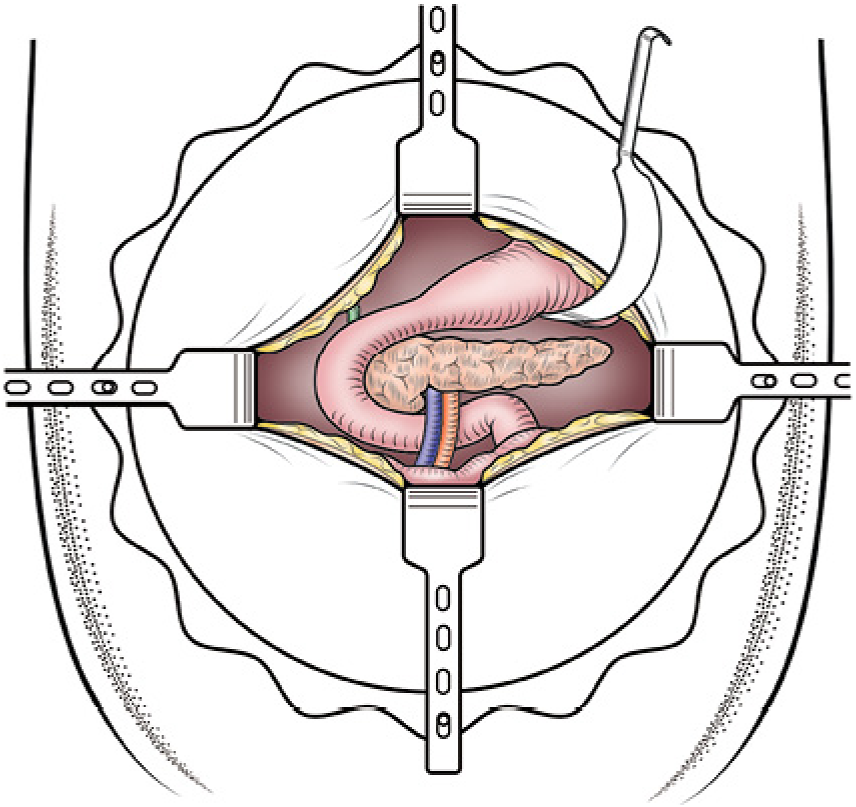
The gold-standard imaging modality in HI is the ^{18}fluoro-L-3-4 dihydroxyphenylalanine positron emission tomography merged with a low-radiation CT scan (18F-DOPA PET/CT) (**Figure 64.3a and b**). Islet cells of the pancreas take up L-DOPA, convert it to L-dopamine through the enzyme L-DOPA decarboxylase, and store it in vesicles. Similarly, these cells can take up ^{18}fluoro-L-3-4 dihydroxyphenylalanine (18F-DOPA), convert it into ^{18}fluoro-dopamine and store it in vesicles that can be traced by their gamma radiation. The study is done under general anesthesia in a PET/CT hybrid scanner that initially captures the radioactive signal and then generates a low-radiation CT scan of the abdomen without moving the patient. Focal lesions are seen as bright spots over a dark background due to the high concentration of the tracer, whereas in diffuse HI the tracer is homogeneously distributed throughout the organ. It is important to note that in about 15% of infants with focal HI nothing can be demonstrated on 18F-DOPA PET/CT.



64.3a,b 18F-DOPA PET/CT scan. (a) Diffuse HI. The 18F-DOPA is distributed evenly throughout the head, body, and tail of the pancreas (black arrows). (b) Focal HI. The 18F-DOPA is taken up by a discrete region in the tail of the pancreas (dotted arrow). The black star marks the left kidney.

OPERATIONS

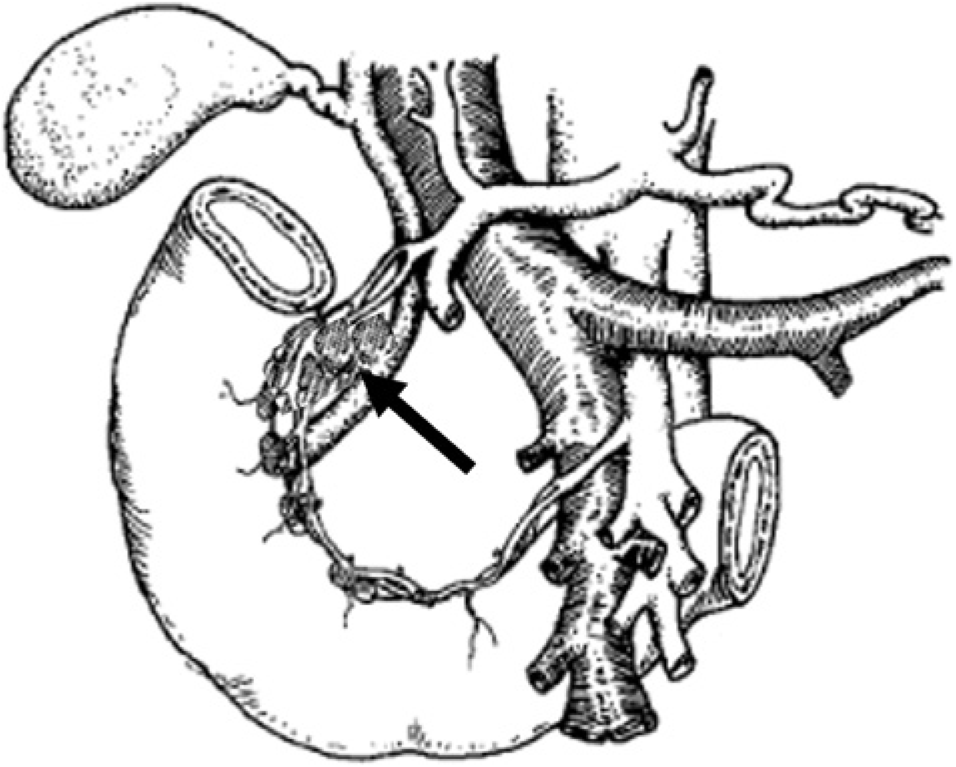
The operation is done through a transverse supraumbilical laparotomy. A Denis Browne retractor provides optimal exposure. The pancreas is completely exposed by mobilizing the hepatic flexure of the colon toward the infant's left side, and an extended Kocher maneuver frees the posterior aspect of the pancreatic head, accessing the lesser sac through the gastrocolic ligament, and retracting the stomach upward with a Deaver retractor (**Figure 64.4**). Regardless of the results of the preoperative genetic testing and imaging studies, the pancreas must be thoroughly inspected under 3.5 × loupe magnification and carefully palpated in an attempt to identify a focal lesion. Focal lesions are often firmer than normal pancreatic tissue and may have subtle differences in appearance and texture. If no obvious focal lesion is identified, small biopsies are taken sharply (cautery is avoided because it hampers frozen section pathology interpretation) from random areas the pancreatic head, body, and tail for intraoperative frozen section analysis. Expert pediatric anatomic pathology interpretation is crucial for a successful operation.



64.4

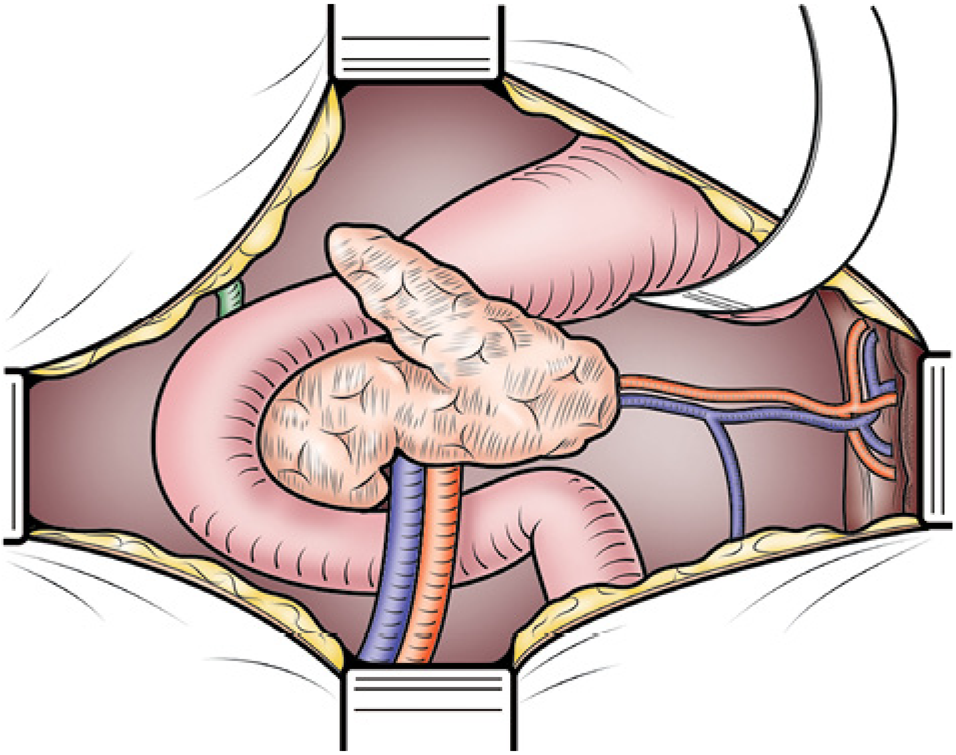
Diffuse hyperinsulinism: Near-total pancreatectomy

A true near-total (98%) pancreatectomy involves the resection of virtually the entire pancreas, leaving only a small residual piece of pancreatic tissue between the common bile duct (CBD) and the duodenal wall, marked by an arrow on **Figure 64.5**.



64.5

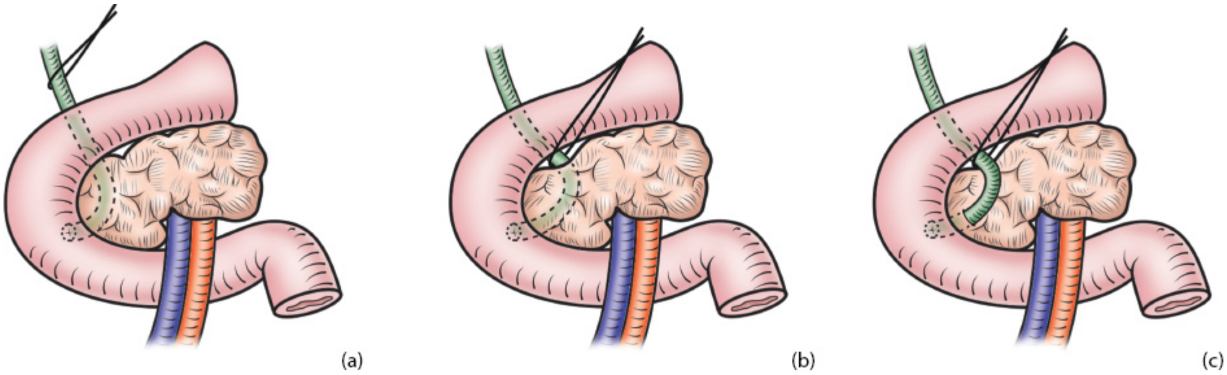
After confirming the diagnosis of diffuse disease by pancreatic biopsies with frozen section, the pancreatectomy starts by carefully dissecting the tail of the pancreas off the splenic hilum (**Figure 64.6**). It is important to clearly distinguish between adipose and pancreatic tissue so that no pancreatic tissue is left behind. The dissection progresses from the tail toward the head. The pancreatic tail and body receive multiple small branches from the splenic artery and vein, which must be carefully cauterized with monopolar or bipolar cautery in order to preserve the splenic artery and vein.



64.6

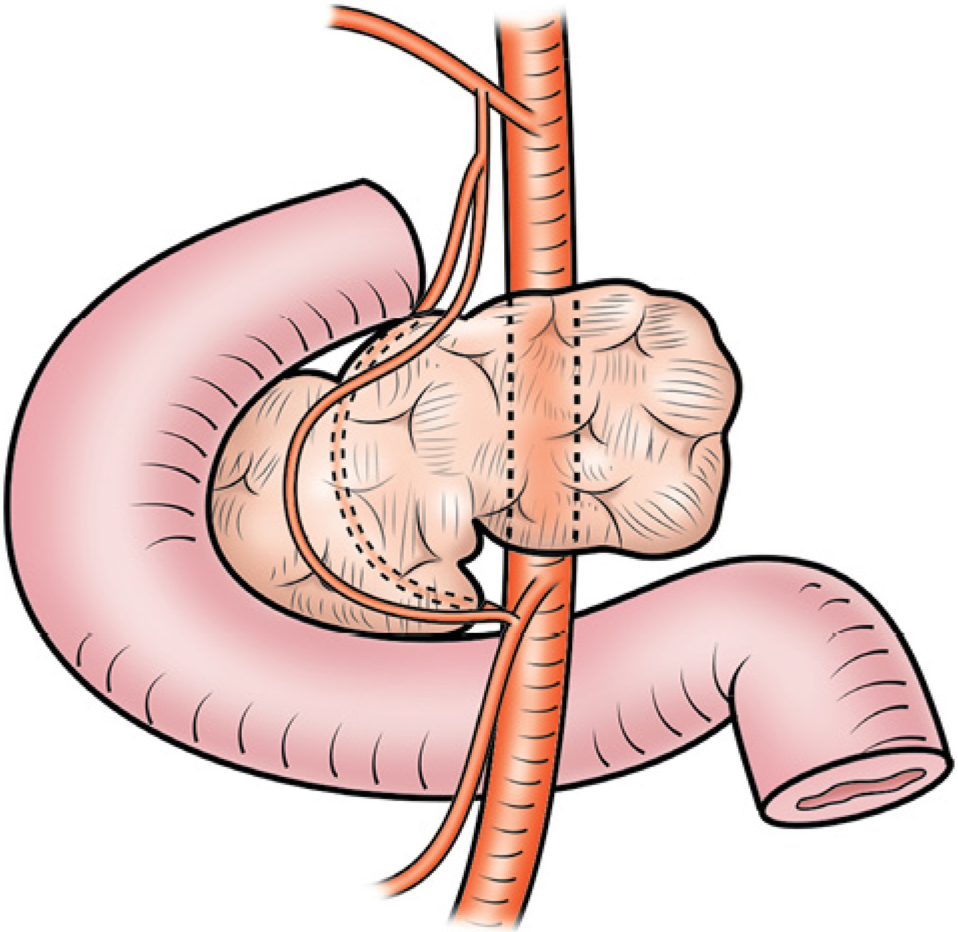
Apart from those small vascular branches, the plane between the posterior aspect of the pancreatic body/tail and the retroperitoneum is relatively avascular. The dissection continues until the pancreatic neck is reached, which is the area overlying the mesenteric vessels. At this point, the confluence between the superior mesenteric vein and the splenic vein giving origin to the portal vein must be clearly identified. Lymphatics are ligated at the inferior aspect of the pancreatic neck to prevent chylous ascites. To help with the exposure of the superior pancreatic edge at the neck/head, the right gastroepiploic artery is ligated just as it comes off the gastroduodenal artery.

The operation continues with the resection of the pancreatic head, which constitutes the more challenging part of the procedure. The key points during this step are first, to preserve the integrity of the intrapancreatic segment of the CBD, and second, to preserve the blood supply to the duodenum by preserving the pancreaticoduodenal arteries. The distal CBD can be completely visible lying over the posterior aspect of the pancreatic head, which makes its dissection relatively easy, or it can be embedded and hidden within the parenchyma of the pancreatic head. In the latter cases, to help with the identification of the hidden segment of the CBD, a vessel loop is placed around the extrapancreatic supraduodenal section of the CBD (**Figure 64.7a**), then a tunnel is made between the first portion of the duodenum and the superior edge of the pancreatic head by careful blunt dissection, and the vessel loop is swung medially toward the duodenal C-loop (**Figure 64.7b**). This allows seeing the CBD on the anterior/superior aspect of the pancreatic head, and carefully skeletonizes it all the way to its insertion in the medial wall of the second portion of the duodenum (**Figure 64.7c**).



64.7a–c A vessel loop is placed around the CBD above the duodenum and swung (dotted arrow) within the concavity of the duodenal C-loop (a). Once identified on the anterior aspect of the pancreatic head (b), it can be safely dissected all the way to its insertion in the duodenum (c).

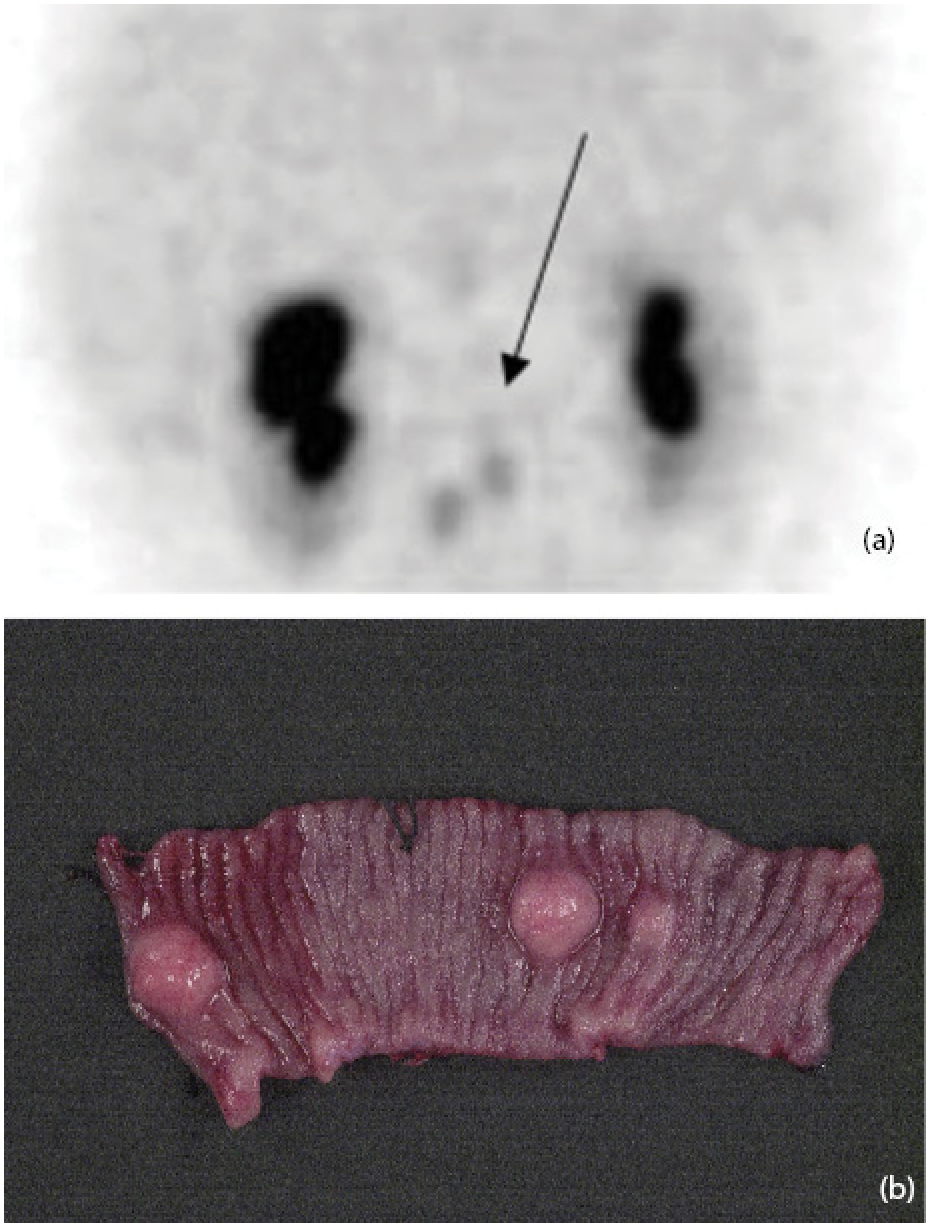
The inferior aspect of the pancreatic head extends medially underneath the superior mesenteric vessels to form the uncinate process (**Figure 64.8**). The dissection of this region must preserve the integrity of the posterior and anterior inferior pancreaticoduodenal arteries, which come off the superior mesenteric artery and provide blood supply to the third and fourth portions of the duodenum. The dissection of the pancreatic head off the proximal and distal duodenum, and off the medial aspect of the CBD completes the near-total pancreatectomy. Generally, the lumen of the proximal cut end of the pancreatic duct is not visible and there is no need to formally close it, but, if there is any concern for a potential leak, the cut end of the remaining pancreatic tissue can be oversewn with interrupted non-absorbable mattress sutures. A drain is not needed after pancreatic resection for HI in infants.



64.8

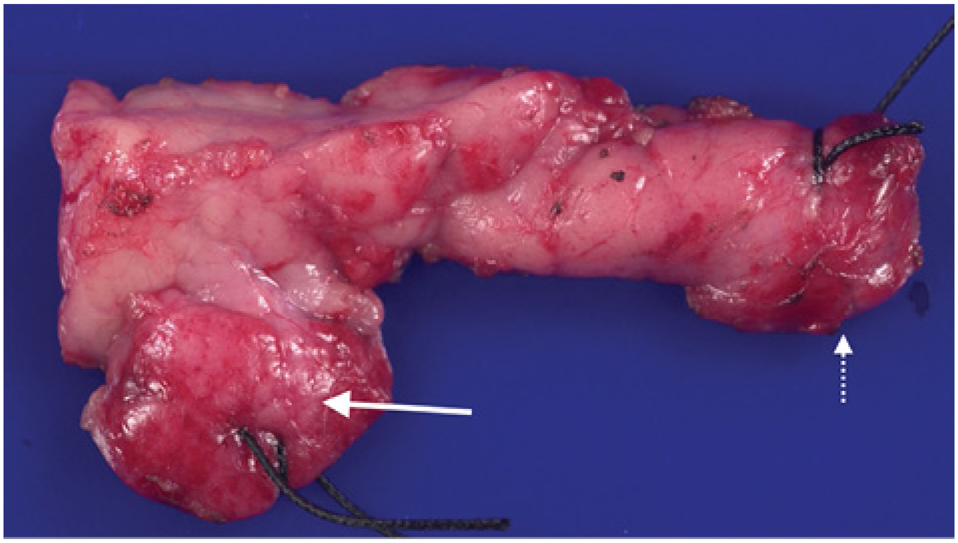
Focal hyperinsulinism: Localized pancreatic resection

If the intraoperative biopsies of the head, body, and tail confirm normal pancreatic histology, a search for the focal lesion is conducted. The preoperative PET/CT should provide invaluable guidance, as it is about 100% accurate in the determination of the location of detected focal lesions, even in the rare cases of ectopic focal lesions (**Figure 64.9a and b**).



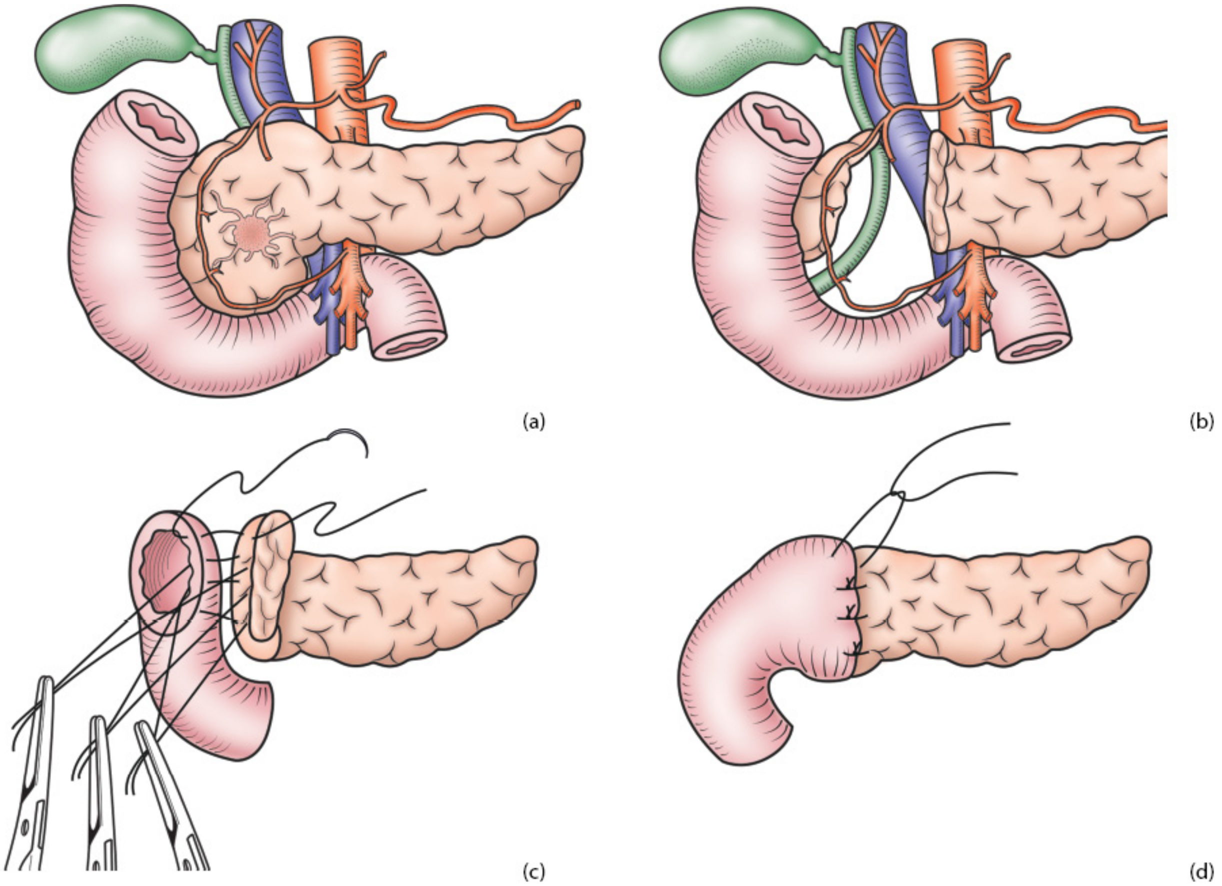
64.9a,b Ectopic focal lesions of HI located in the small bowel, accurately detected by PET (a) and CT (b).

Intraoperative high-resolution US is usually not helpful in localizing focal lesions but US can delineate the course of the pancreatic duct, which is about 0.4 mm in diameter. Intraoperative US may be particularly helpful when the genetic testing suggests a focal lesion but the PET/CT does not show it (in about 15% of cases). Focal lesions that are deep within the pancreatic tissue are impossible to see or feel by palpation, so it is necessary to patiently take biopsies of suspicious areas for frozen section analysis until the lesion is found. Focal lesions are irregularly shaped and frequently have octopus-like tentacles, which makes the intraoperative frozen section confirmation of clear margins imperative. Once the focal lesion is identified, a partial pancreatectomy is performed (**Figure 64.10**). Disease-free margins must be confirmed before concluding the surgery. Small and superficial lesions in the body or tail are treated by local resection. Large and/or deep lesions in the body and tail are treated by distal pancreatectomy.



64.10 Superficial focal lesion in the pancreatic body (white arrow). The lesion has a different color than the normal pancreatic tissue. The dotted arrow marks the pancreatic tail.

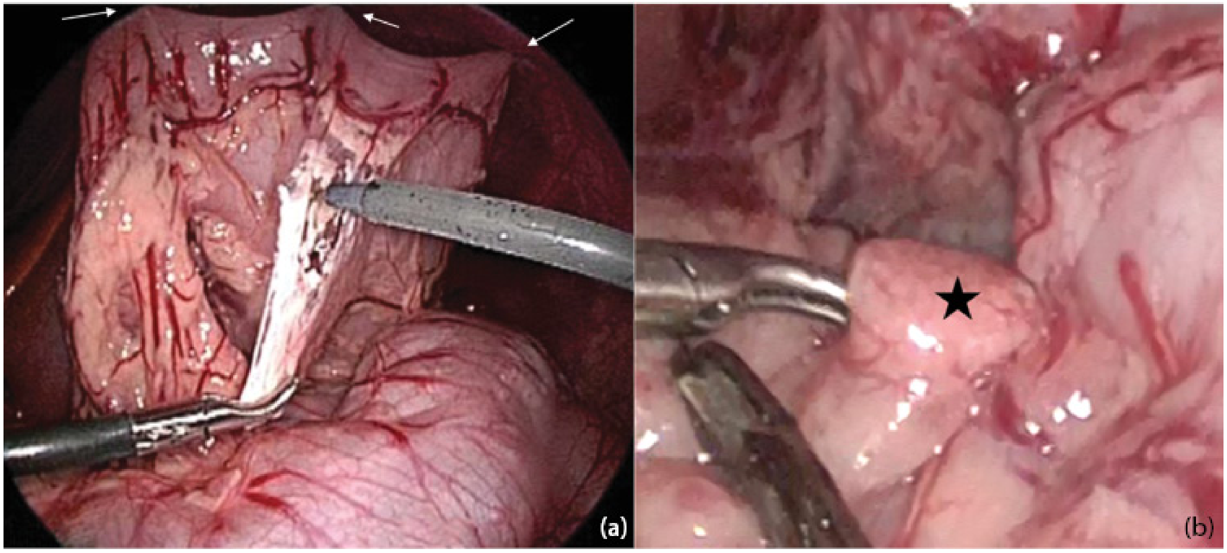
In the case of a distal pancreatectomy, to prevent leakage from the pancreatic duct, a transfixion suture of 2/0 silk is placed and tied just proximal to the area through which the pancreas will be transected. After the transection and once the distal pancreatectomy specimen is removed, an additional mattress suture of 3/0 silk is placed and tied. Superficial and small lesions in the head of the pancreas can also be treated by local resection. On the other hand, deep lesions of the pancreatic head can be difficult to excise with clear margins without risking damage to the CBD or pancreatic duct. To ensure a complete resection of the lesion in these challenging cases, almost the entire pancreatic head can be resected, followed by a Roux-en-Y pancreaticojejunostomy to drain the remaining pancreatic body and tail (**Figure 64.11a–c**). This approach is required in about 40% of focal lesions located in the pancreatic head. The end of a retrocolic 25 cm-long Roux-en-Y jejunal limb is meticulously anastomosed to the capsule of the pancreatic body (just beyond the cut end of the pancreas) with fine interrupted 5/0 monofilament sutures to tuck the cut end of the pancreas into the jejunal lumen. The posterior aspect of the anastomosis is performed first, with all sutures placed and then tied serially, leaving the knots on the inside of the anastomosis. The anterior aspect is performed in the same manner, but leaving the knots on the outside. The omentum is then wrapped around the anastomosis to help prevent an anastomotic leak. Rarely, a focal lesion in the head will extend into the duodenal wall, in which case a Whipple procedure may be needed.



64.11a–c (a) Focal lesion in the head of the pancreas that has octopus-like tentacles that extend into the normal tissue. (b) Near-total pancreatic head resection. The common bile duct is skeletonized and the duodenal vasculature is preserved. A very small portion of the pancreatic head (<5%) is left between the CBD and the duodenal wall, along the superior pancreaticoduodenal artery. (c) Pancreaticojejunostomy.

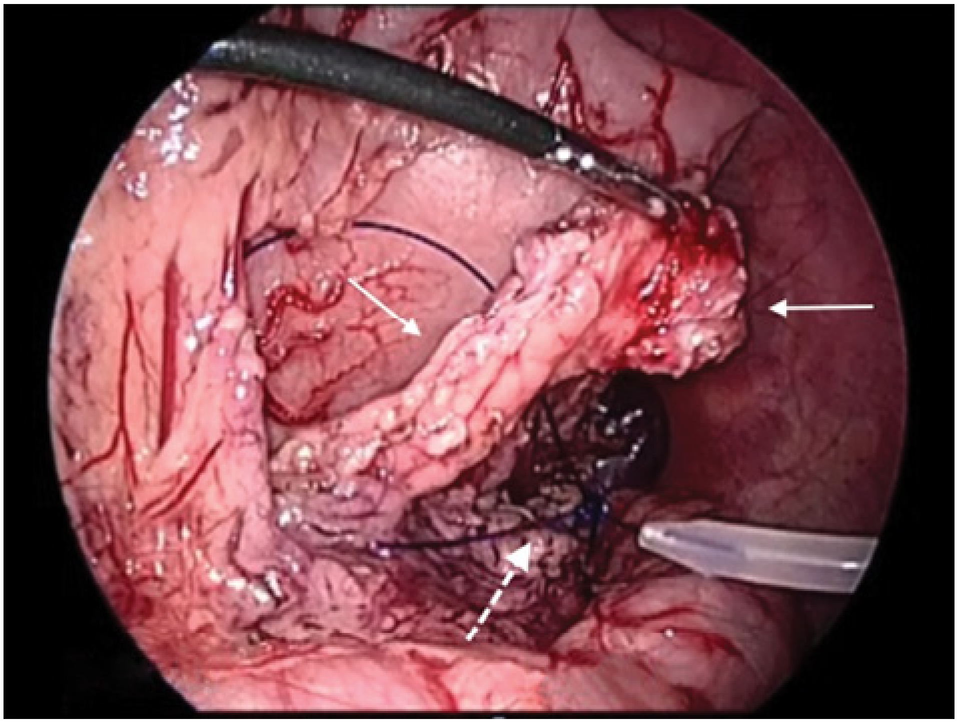
Laparoscopic approach

Laparoscopic surgery is sometimes suitable in HI patients with focal disease of the pancreatic body or tail. The surgery is done via three or four 3–5 mm ports and to facilitate pancreatic exposure the stomach is tacked up to the anterior abdominal wall with two or three transabdominal–transgastric stitches near the greater curvature (Figure 64.12a). Superficial focal lesions located in the anterior pancreatic aspect are relatively easy to detect laparoscopically (Figure 64.12b).



64.12a,b (a) Laparoscopic exposure of the pancreas. The stomach is retracted upward and held by several trans-gastric, trans-parietal stitches (white arrows). The gastrocolic ligament is taken down with cautery. (b) A focal lesion (black star) is seen on the anterior aspect of the pancreatic body.

The laparoscopic approach has two limitations: Tactile feedback is not available for the palpation of deep non-visible focal lesions, and intraoperative ultrasound cannot be performed adequately via a small laparoscopic port to evaluate the location of the pancreatic duct. The dissection and resection of the pancreatic tail and body for a distal pancreatectomy is relatively straightforward (**Figure 64.13**). The small vessels that arise from the splenic artery and vein are carefully cauterized with monopolar current, and the pancreas is ligated proximally to the desired area of transection with one or two loop ties.



64.13 Laparoscopic distal pancreatectomy. The pancreatic body and tail (white arrows) have been dissected off the splenic vessels. A loop tie is about to be placed proximal to the area of the planned transection (white dotted arrow).

The dissection and resection of the pancreatic head is much more technically demanding than the dissection of the distal pancreas. A high rate of CBD injury has been observed in cases of laparoscopic near-total pancreatectomy to treat diffuse disease. Literature reports that claim a low rate of CBD injuries consist of cases in which the CBD is neither identified nor dissected and the pancreatectomy is carried just beyond the superior mesenteric vessels, are not near-total but rather subtotal or distal pancreatectomies with an attendant risk of persistent postoperative hypoglycemia.

POSTOPERATIVE CARE

Postoperative pain after open pancreatectomy is best managed by an epidural catheter placed prior to the operation, and IV narcotics. The IV glucose infusion rate (GIR) is restarted immediately after the operation at a very low rate (2 mg/kg/min) in part because the stress of the surgery induces hepatic glycogenolysis. GIR is advanced to 5 mg/kg/min 12–18 hours after the surgery and to 8 mg/kg/min (which is equivalent to the physiological hepatic glucose release rate during fasting periods) 24–36 hours after the surgery. Plasma glucose levels are measured hourly initially and spaced out as they become stable. If the plasma glucose levels are excessively high (>400 mg/dL, ≈22 mmol/L) IV insulin infusion is started. The immediate postoperative oscillations in the plasma glucose levels do not reflect the eventual long-term outcome because surgical stress and postoperative pain alter glucose homeostasis.

When bowel function resumes, enteral feedings are advanced gradually and the GIR is simultaneously weaned down. Once patients are exclusively on enteral feeds, a “cure” fasting test is performed. If the patient is able to maintain euglycemia for ≥18 hours, they are considered to be cured. If the time to hypoglycemia is <18 hours, the next step is to determine a regimen of frequent feeds and short fasting periods that will allow the patient to be managed safely at home. Patients who are unable to be weaned from the IV GIR are not cured and need further assessment to determine if additional surgery is required.

COMPLICATIONS

Surgical complications are rare after pancreatic surgery for HI. General postoperative complications are bowel obstruction due to adhesions and small intestinal intussusception. Specific complications of pancreatic surgery such as chylous ascites and pancreatic leaks are very rare in infants. In our experience of about 210 near-total pancreatectomies, 40 pancreatic head resections/Roux-en-Y reconstructions, and more than 100 resections of focal lesions in the pancreatic head, CBD complications (intraoperative injury or postoperative stricture) occurred in only six cases and were treated with a choledochoduodenostomy.

OUTCOME

In our experience with more than 240 patients with focal HI, more than 97% are cured after the resection of the focal lesion. Failure to achieve a cure is always the consequence of an incomplete resection, which can generally be confirmed by pathology. Patients in that situation may need a redo operation. Nearly 3% of the cases in our experience failed to achieve a cure despite disease-free margins on the permanent pathology analysis. These cases were either very large focal lesions or focal lesions in the pancreatic head with presumed microscopic residual focal disease within the duodenal wall. This microscopic residual disease burns out over time.

The outcome of infants with diffuse HI who undergo a near-total pancreatectomy is less predictable. About 30% of patients achieve normoglycemia soon after the operation and do not require any additional hypoglycemic therapy. About 50% continue to have hypoglycemia after surgery and require supportive management with frequent or even continuous feeds. Despite this less-than-ideal outcome, these patients are much more easily manageable after surgery than before the surgery and are at home off IV glucose infusions. The remaining 20% of patients develop early diabetes requiring insulin. Recent studies have shown that the incidence of insulin-dependent diabetes increases with time, to reach more than 90% 10 years after the operation. The long-term incidence of clinically evident exocrine insufficiency is also high at about 50%. Additionally, neurodevelopmental abnormalities are relatively common in patients with HI, even in those who have not experienced severe episodes of hypoglycemia.

VIDEOS

OPS_8e_video_64.1 Laparoscopic pancreatectomy

FURTHER READING

Adzick NS, Thornton PS, Stanley CA, et al. A multidisciplinary approach to the focal form of congenital hyperinsulinism leads to successful treatment by partial pancreatectomy. *J Pediatr Surg* 2004; 39: 270–5.

Adzick NS, DeLeon DD, States LJ, et al. Surgical treatment of Congenital Hyperinsulinism: Results from 500 pancreatectomies in neonates and children. *J Pediatr Surg* 2019; 54:27–32.

Laje P, Stanley CA, Palladino AA, et al. Pancreatic head resection and Roux-en-Y pancreaticojejunostomy for the treatment of the focal form of congenital hyperinsulinism. *J Pediatr Surg* 2012; 47: 130–5.
Laje P, States LJ, Zhuang H, et al. Accuracy of PET/CT scan in the diagnosis of the focal form of congenital hyperinsulinism. *J Pediatr Surg* 2013; 48: 388–93.
Lord K, Radcliffe J, Gallagher PR, et al. High risk of diabetes and neurobehavioral deficits in individuals with surgically treated hyperinsulinism. *J Clin Endocrinol Metab* 2015; 100: 4133–9.
Peranteau WH, Bathaii SM, Pawel B, et al. Multiple ectopic lesions of focal islet adenomatosis identified by positron emission tomography scan in an infant with congenital hyperinsulinism. *J Pediatr Surg* 2007; 42: 188–92.
Stanley CA. Perspective on the genetics and diagnosis of congenital hyperinsulinism disorders. *J Clin Endocrinol Metab* 2016; 101: 815–26.

65

Adrenalectomy

MIKAEL PETROSYAN AND TIMOTHY D. KANE

HISTORY

The first recorded adrenalectomy involved the en bloc removal of a 9 kg tumor from a 36-year-old woman with hirsutism and virilizing features (most probably a malignant adrenocortical carcinoma [ACC]) by Thornton in 1889. Nevertheless, malignant adrenocortical disease is sporadic, accounting for less than 0.2% of all pediatric tumors and only 6% of adrenal tumors. Neuroblastoma is the most common adrenal lesion, accounting for a 50-fold greater number of adrenal tumors than the second most common tumor, pheochromocytoma.

EMBRYOLOGY

The adrenal gland is composed of two types of endocrine tissues of different embryonic origin, the medulla and the cortex. The chromaffin cells of the adrenal medulla are derived from neuroectoderm, whereas the cells of the adrenal cortex are derived from mesoderm. Gonadal tissues arise from a mesodermal source, and since they are similarly involved in the synthesis of steroid hormones, inborn enzymatic defects at one site are frequently present in the other. The common origin may therefore result in both medical and surgical conditions, which involve multiple systems and require multidisciplinary approaches to treatment.

The adrenal cortex may be divided into three zones:

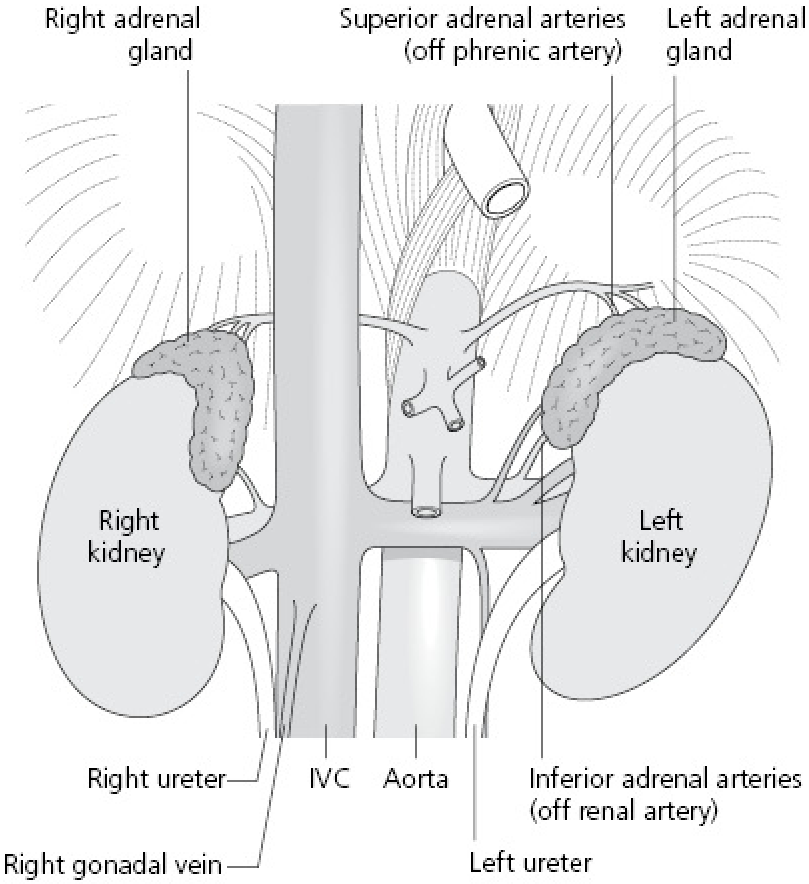
- The *zona glomerulosa* is the outermost zone of the cortex, responsible for the synthesis of aldosterone.
- The *zona fasciculata* is the largest zone of the cortex and the site of cortisol production.
- The *zona reticularis* is the innermost and smallest zone. Cells here produce adrenal androgens, dihydroepiandrosterone, and androstenedione.

The adrenal medulla is composed of neuroendocrine (chromaffin) and glial cells.

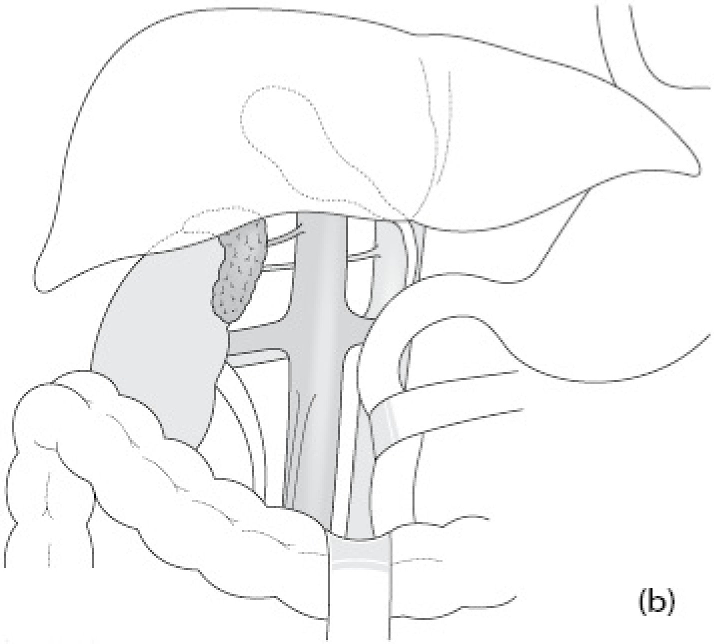
Development of the adrenal gland begins between 3 and 4 weeks of gestation and cephalad to the developing mesonephros. At this point, the gonadal ridge has yet to form, and there is no distinction between adrenal and gonadal tissue. By 5–6 weeks, however, steroidogenic gonadal cells begin their caudal migration, while the cells that will eventually comprise the zona reticularis migrate dorsally into the retroperitoneum. Over the next several weeks, there is a rapid enlargement of the inner cortex to form the fetal zone (the outer subcapsular rim remains as the definitive zone) and, simultaneously, migrating cells of the neuroectoderm also follow trophic signals to populate the adrenal medulla. These cells will subsequently be responsible for producing and storing catecholamines. The fetal adrenal gland is proportionally much larger than that in the adult. At 8 weeks' gestation, the adrenals are larger than the kidney, and at term, they are approximately one-third the size of the kidney, with the fetal cortex comprising most of that mass. Within several days of birth, the fetal cortex begins rapid involution and decreases to half the immediate postnatal size at 1 month and one-eighth of its size by 1 year. During this time, the medulla begins a slow period of growth. The relative size of the neonatal adrenal gland is thought to contribute to perinatal adrenal hemorrhage (reported incidence three in 100 000 live births). Occasionally, adrenal hemorrhage may lead to exsanguination and death, or present with an abdominal mass, anemia, or scrotal hematoma.

ANATOMY

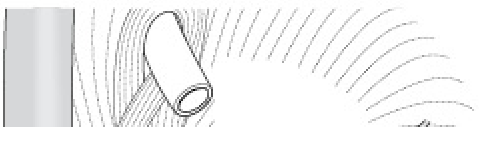
The adrenal glands are paired but not identical structures that lie superomedially to the kidneys at the level of the eleventh thoracic vertebrae. Posteriorly, both glands abut the posterior aspect of their respective hemidiaphragms. The posterior right lobe of the liver borders the superior surface of the right adrenal, and the inferior vena cava (IVC) borders the medial aspect of the adrenal gland. The right gland is triangular, slightly smaller, and positioned more cephalad than the left. The left adrenal gland is an elongated, flat structure that is situated posterior to the splenic vessels and the tail of the pancreas (**Figure 65.1a–c**).

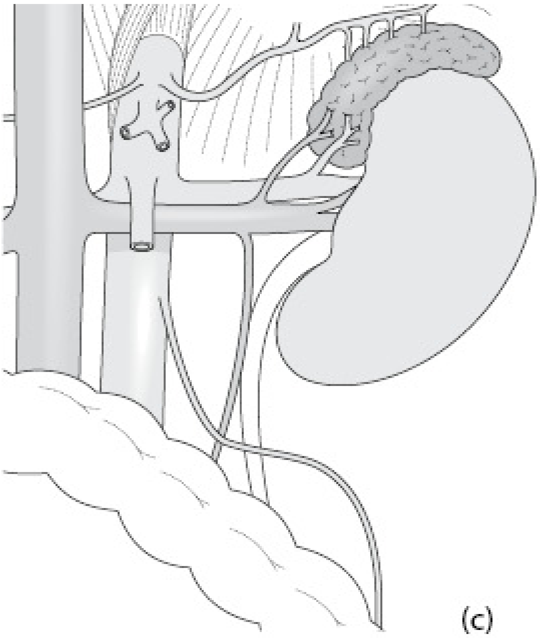

Right adrenal gland
Superior adrenal arteries (off phrenic artery)
Left adrenal gland
Right kidney
Left kidney
Right ureter
IVC
Aorta
Inferior adrenal arteries (off renal artery)
Right gonadal vein
Left ureter

(a)



(b)





65.1a–c

The adrenal glands receive arterial blood supply from three main sources: branches of the phrenic artery from the celiac axis, direct aortic perforating vessels, and small perforating vessels from the renal artery. In fact, there is rarely a distinct adrenal artery. Instead, an end vascular arcade from those three sources supplies the blood to the adrenal glands. During surgical resection, caution must be used to adequately cauterize these arterial sources to minimize intraoperative and postoperative blood loss. In contrast, venous drainage of the adrenal glands reliably occurs through a dominant vein. On the right side, this vein empties directly into the posterolateral IVC, 3–4 cm above the right renal vein, although there may be a smaller accessory right adrenal vein. The left adrenal vein drains directly into the left renal vein.

During embryogenesis, primordial adrenal cells may migrate caudally with other cells of the gonadal ridge. This migration accounts for the presence of accessory adrenal tissue in the retroperitoneum along the course of the descent of the ovaries and testes.

OPERATION

Conditions requiring adrenalectomy include both benign and malignant disease. Benign conditions include enlarging or endocrinologically active adrenal adenomas and hormone- or catecholamine-secreting neoplasms, including pheochromocytoma and aldosteronoma. Malignant conditions include neuroblastoma, ganglioneuroblastoma, ACC, and metastatic lesions.

A variety of operative approaches exist and selection of any particular approach must be individualized according to the adrenal pathology and mass size, patient body habitus, previous operative intervention, and surgeon experience. Traditional approaches to adrenal surgery have included anterior, posterior, and thoracoabdominal techniques. The transabdominal approach is appropriate for large or bilateral tumors, tumors encasing central vascular structures (e.g. neuroblastoma), or for large tumors such as pheochromocytoma, which may also be found in extra-adrenal locations. A complete abdominal exploration may be performed via this approach. The posterior approach is appropriate for resection of small adenomas or aldosteronomas, or in patients in whom previous abdominal surgery has rendered the abdomen hostile to further operative approaches. A posterior approach includes the additional advantage of preventing the development of future abdominal adhesions and reduces the likelihood of postoperative small bowel obstruction. It is also associated with a shorter functional recovery and shorter hospital stay. Disadvantages include the impracticality of examining the abdominal viscera or contralateral adrenal as well as the relatively limited exposure in the event of vascular injury.

Over the last decade, increasing experience has accumulated with laparoscopic adrenalectomy such that it accounts for greater than 75% of adrenal resections in some centers. Ideal candidates for a laparoscopic approach are those patients with small, well-circumscribed, benign-appearing lesions. Although some authors have reported the successful laparoscopic removal of ACC, most surgical oncology authorities consider ACC a contraindication to the minimally invasive approach. Not only does the tumor have the tendency to invade nearby structures, but also capsular rupture will upstage the disease.

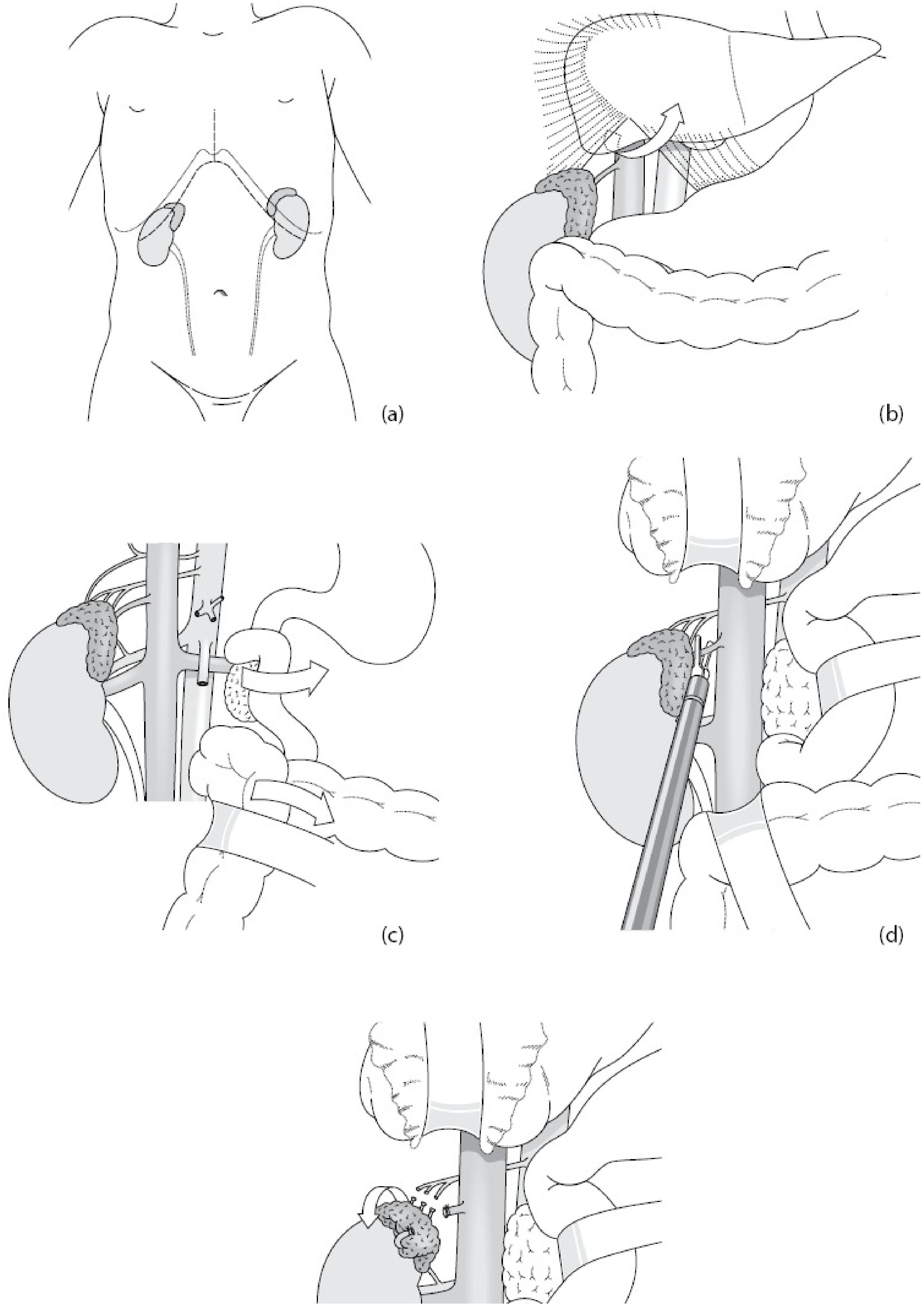
Anterior approach

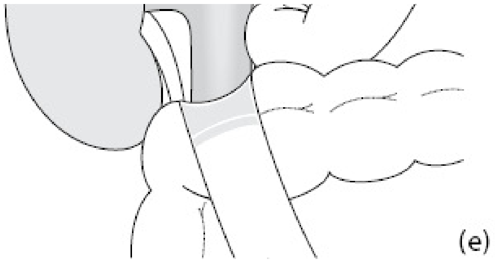
The anterior approach provides excellent bilateral exposure to the structures of the upper abdomen and is frequently the approach of choice for large, malignant, or bilateral lesions. In very large or obese patients, intra-abdominal fat and the position of the adrenal glands is higher under the dome of the diaphragm, thus a thoracoabdominal approach may be most appropriate. Postoperative morbidity is greater than with a posterior or laparoscopic approach with an increased risk of adynamic ileus, atelectasis, and incisional discomfort.

RIGHT ADRENALECTOMY

The child should be positioned supine on the operating room table with a roll under the right lower costal margin. A subcostal incision is typically employed and can be extended across the midline (chevron incision) for better exposure (**Figure 65.2a**); however, a midline incision may also be used. Upon entering the abdomen, the hepatic flexure of the colon is reflected inferomedially, and a wide Kocher maneuver is performed, exposing the IVC. An external retractor (Buchwalter, Thompson, or Omni) is helpful to maintain exposure. The right lobe of the liver should be gently retracted cephalad to expose the superior surface of the adrenal gland that should be palpable superomedial to the kidney and lateral to the IVC (**Figure 65.2b**). Occasionally, a tongue of adrenal can extend posteriorly to the IVC. The peritoneum overlying the adrenal should be fully exposed. The lateral border can be mobilized by the application of serial clamps and ties or by electrocautery (**Figure 65.2c**). As the arterial supply to the adrenal comes from small perforating branches from the phrenic artery, aorta, and renal artery, the lateral border is relatively avascular. The medial border should then be dissected free of the IVC. Often, this will require a near-complete mobilization of the posterior surface of the IVC, allowing it to be retracted gently to the left. The right adrenal vein is a short, wide vessel that drains directly into the posterolateral IVC (**Figure 65.2d**). The vein can be easily avulsed resulting in significant bleeding and gentle, careful dissection should be used during its dissection. Once the right adrenal vein is mobilized, it should be ligated and divided (**Figure 65.2e**).

Many surgeons place a clip behind the tie on the IVC for additional security. Inadvertent holes in the IVC should be repaired with 5/0 Prolene sutures. Urgent control of the IVC may be obtained if necessary by the judicious use of sponge sticks or a side-biting Satinsky clamp. The inferior and posterior surfaces of the adrenal gland are relatively avascular and dissection from the kidney and retroperitoneum may be accomplished with the use of electrocautery. The adrenal tissue itself may be quite friable and care should be taken during its dissection to avoid too much traction on the tissue, which may result in organ fragmentation and bleeding.

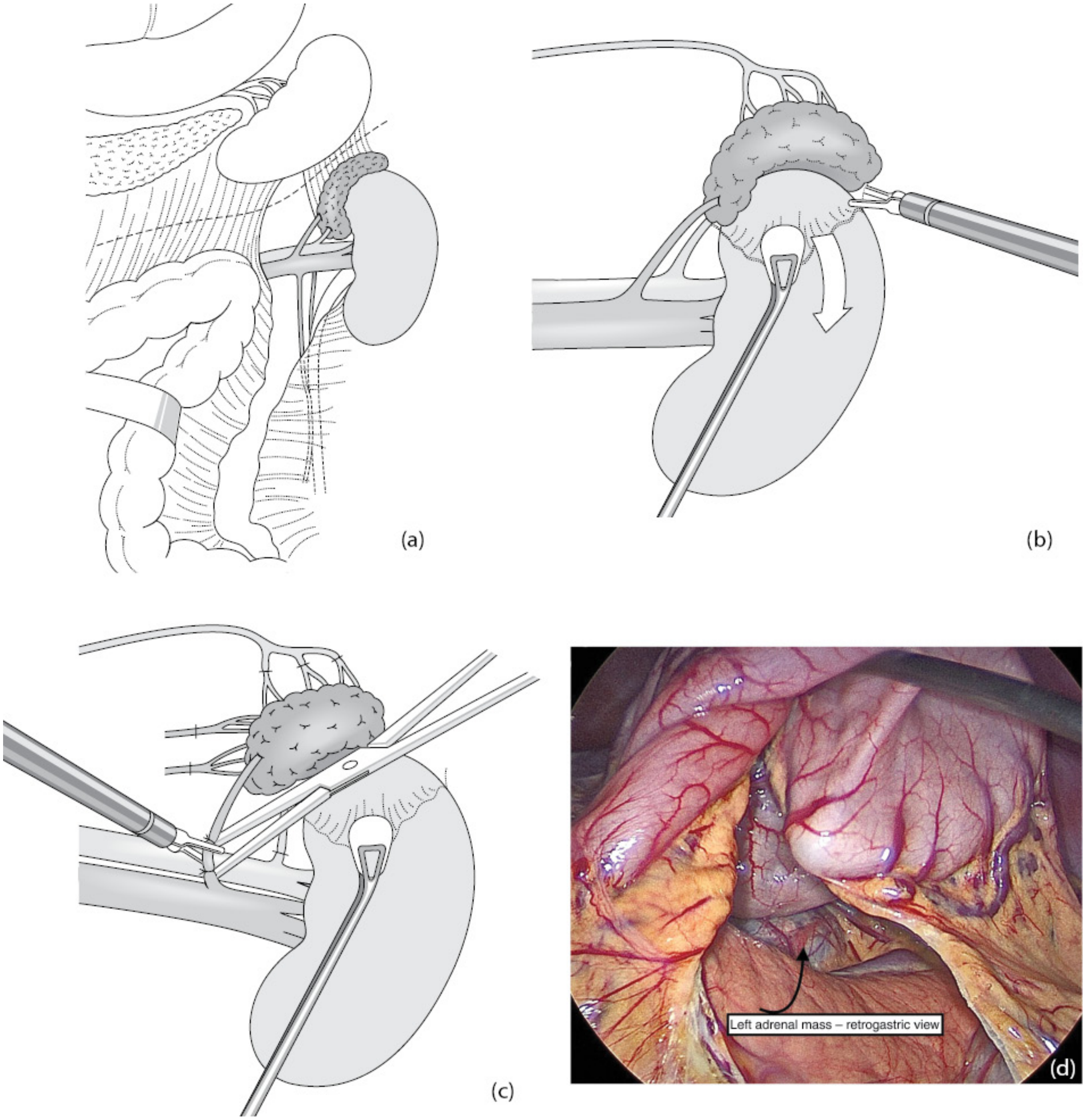

(a)
(b)
(c)
(d)



65.2a–e

LEFT ADRENALECTOMY

The patient is positioned in a similar fashion to that for right adrenalectomy with a roll under the lower left costal margin. Access to the left adrenal gland may be obtained either through inferior reflection of the left colon and splenic flexure or via the lesser sac (retrogastric approach). In this approach, the gastrocolic omentum is widely divided, taking care to preserve the gastroepiploic arcade supplying the greater curvature of the stomach (**Figure 65.3a**). Upon entry into the lesser sac, the stomach is reflected superiorly and the peritoneal surface overlying the inferior border of the distal pancreas is incised, allowing the pancreas to be gently retracted caudad (**Figure 65.3b**). Downward retraction of the transverse colon typically allows visualization of the left renal vein, which may be traced laterally to identify the medial border of the kidney and adrenal. The lienorenal ligament should be divided. Often, this approach requires more complete mobilization of the splenic flexure. Gerota's fascia overlying the superomedial border of the kidney should be incised to expose the adrenal gland, which has a bright yellow, bosselated, and slightly grainy appearance. The anterior surface of the left renal vein should be dissected. The gonadal vein, which enters the inferior edge of the left renal vein, is a helpful landmark. The left adrenal vein enters the renal vein opposite the gonadal on the superior surface, typically about a centimeter closer to the renal hilum. Mobilization of the adrenal begins by the division of small arterial branches (predominantly supplied by the left phrenic artery) on the medial and superior surface with clips, ties, or energy device (Harmonic scalpel™ [Ethicon Inc., Cincinnati, OH, USA], LigaSure™ [Valleylab, Boulder, CO, USA], etc). The left adrenal vein may now be divided and the stump used for traction to dissect the relatively avascular posterior and inferior surfaces (**Figure 65.3c**).


(a)
(b)
(c)
Left adrenal mass – retrogastric view
(d)

65.3a–d

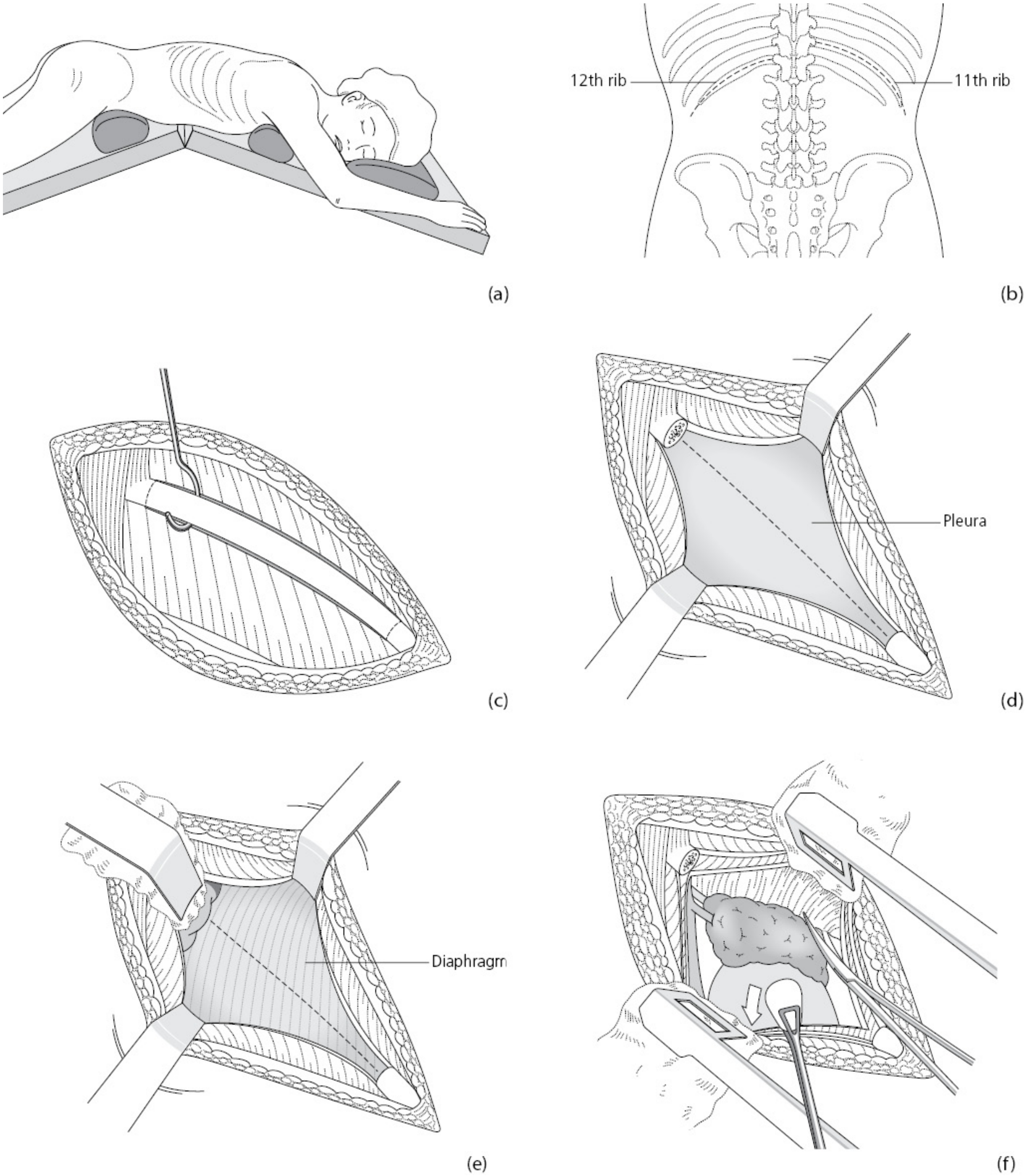
A left adrenal mass visualized in the laparoscopic approach is shown in **Figure 65.3d**.

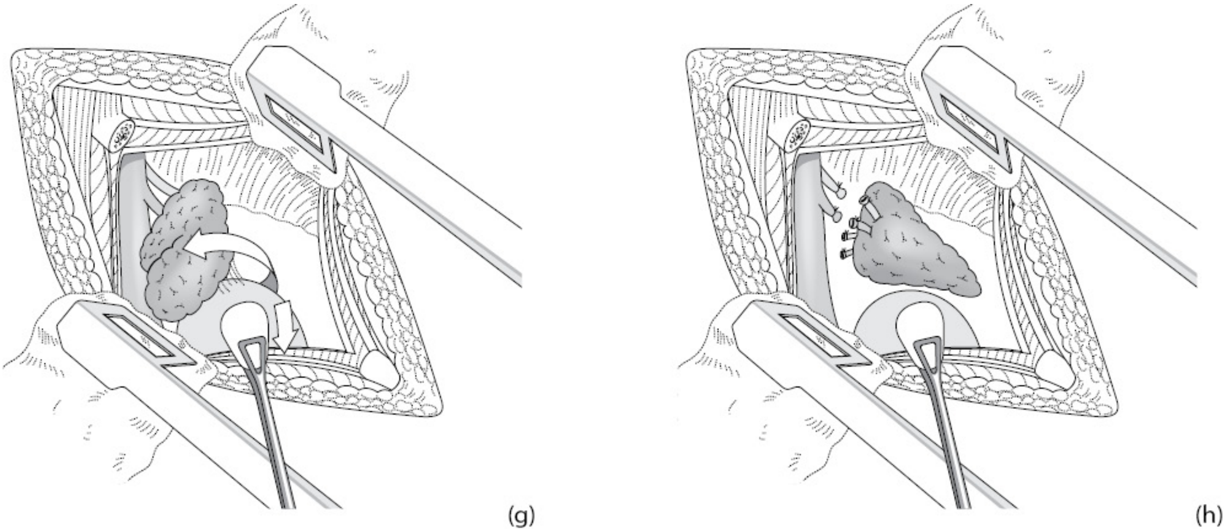
Given the frequency of extra-adrenal disease associated with some adrenal tumors (i.e. pheochromocytoma), the remainder of the retroperitoneum should be palpated when performing an adrenalectomy. This includes palpation of the para-aortic chain and the organ of Zuckerkandl at the aortic bifurcation. Additionally, the bladder should also be checked for masses.

Posterior approach

Young described the posterior approach to adrenalectomy in 1936. The patient is positioned prone on the operating room table with rolls underneath both the chest and hips sufficient to elevate the abdominal wall off the table and allowing the vena cava to fall forward away from the retroperitoneal organs and making the dissection more facile. The table should be angulated (jack-knifed) slightly at the midway point between the anterior superior iliac spine (ASIS) and the costal margin to straighten the curvature of the spine (**Figure 65.4a**). The earliest surgical description featured a "hockey stick" incision that extended vertically downward from the tenth rib, paralleling the spine, before curving gently toward the ASIS, approximately 2 cm below the costal margin. This incision was then carried through the fascia and the inferior fibers of the trapezius and latissimus dorsi muscle. The attachments of the erector spinae muscle to the twelfth rib are divided, the rib resected, and the lumbodorsal fascia is divided longitudinally along the lateral margin of the quadratus lumborum to expose the diaphragm. Division of the diaphragm exposes Gerota's fascia.

This incision has been modified and largely replaced by an incision over the bed of the tenth or eleventh rib on the right or the eleventh or twelfth rib on the left (**Figure 65.4b**). The rib is then resected in the subperiosteal plane as far medial as possible to allow maximal visualization of the adrenal (**Figure 65.4c**). The supraspinatus muscle defines the medial border of the resection and further visualization may be achieved by medial retraction of the muscle. The underlying pleura is divided in the direction of the rib bed (**Figure 65.4d**) and the lung is then retracted medially with a sponge or padded retractor. Next, the diaphragm is divided, exposing the retroperitoneal space (**Figure 65.4e**), and Gerota's fascia is opened superiorly bringing the adrenal into view. Gentle downward traction on the kidney using a sponge stick improves exposure and the placement of a Finochetto retractor frees the assistant's hands for help with dissection (**Figure 65.4f**).


12th rib
11th rib
(a)
(b)
(c)
Pleura
(d)
Diaphragm
(e)
(f)



65.4a–h

On the right side, the liver is gently retracted superiorly (**Figure 65.4g**). Loose adventitial tissue between the adrenal and liver can be carefully dissected. The superior edge of the adrenal is supplied by perforating vessels from the aorta and phrenic artery travelling in an ill-defined vascular “ligament.” This may be divided by a series of clips and electrocautery or by using energy devices such as the Harmonic scalpel™ (Ethicon Inc., Cincinnati, OH, USA) or LigaSure™ (Valleylab, Boulder, CO, USA). The lateral and inferior aspects of the adrenal are relatively avascular and may often be mobilized with monopolar electrocautery alone. The gland is then rotated medially to expose its anatomic anterior surface and facilitate dissection to its medial caval border and it is then returned to the standard position to expose the medial vascular attachments, most notably the adrenal vein. This is ligated and divided prior to completing the dissection of arterial perforating vessels (**Figure 65.4h**).

On the left side, dissection also begins with dissection of the superior vascular perforating vessels and may require gentle upward retraction of the pancreatic tail. The lateral borders are freed (these are largely avascular) and the gland is rotated medially to allow dissection of its anterior (avascular) surface. The adrenal vein may now be identified travelling anterior and medial to the upper pole of the kidney where it joins the left renal vein. This is suture ligated and divided, typically exposing the inferior adrenal artery that is also controlled and divided. The final step involves division of the middle adrenal arteries as they enter the medial surface of the adrenal gland.

A modification of the posterior approach involves a flank approach through the bed of the eleventh or twelfth rib. In this event, the patient is not positioned prone, but in the lateral decubitus position with the hips elevated and the shoulder and hip rotated anteriorly. Subperiosteal resection of the eleventh rib allows for a blunt extrapleural dissection in the posterior rib bed. The diaphragm should be divided at the insertion near the lumbocostal arch. The peritoneum is visualized and reflected anteromedially, exposing the kidney and adrenal. This approach allows excellent exposure of the IVC and the insertion of the adrenal vein on its posteromedial aspect.

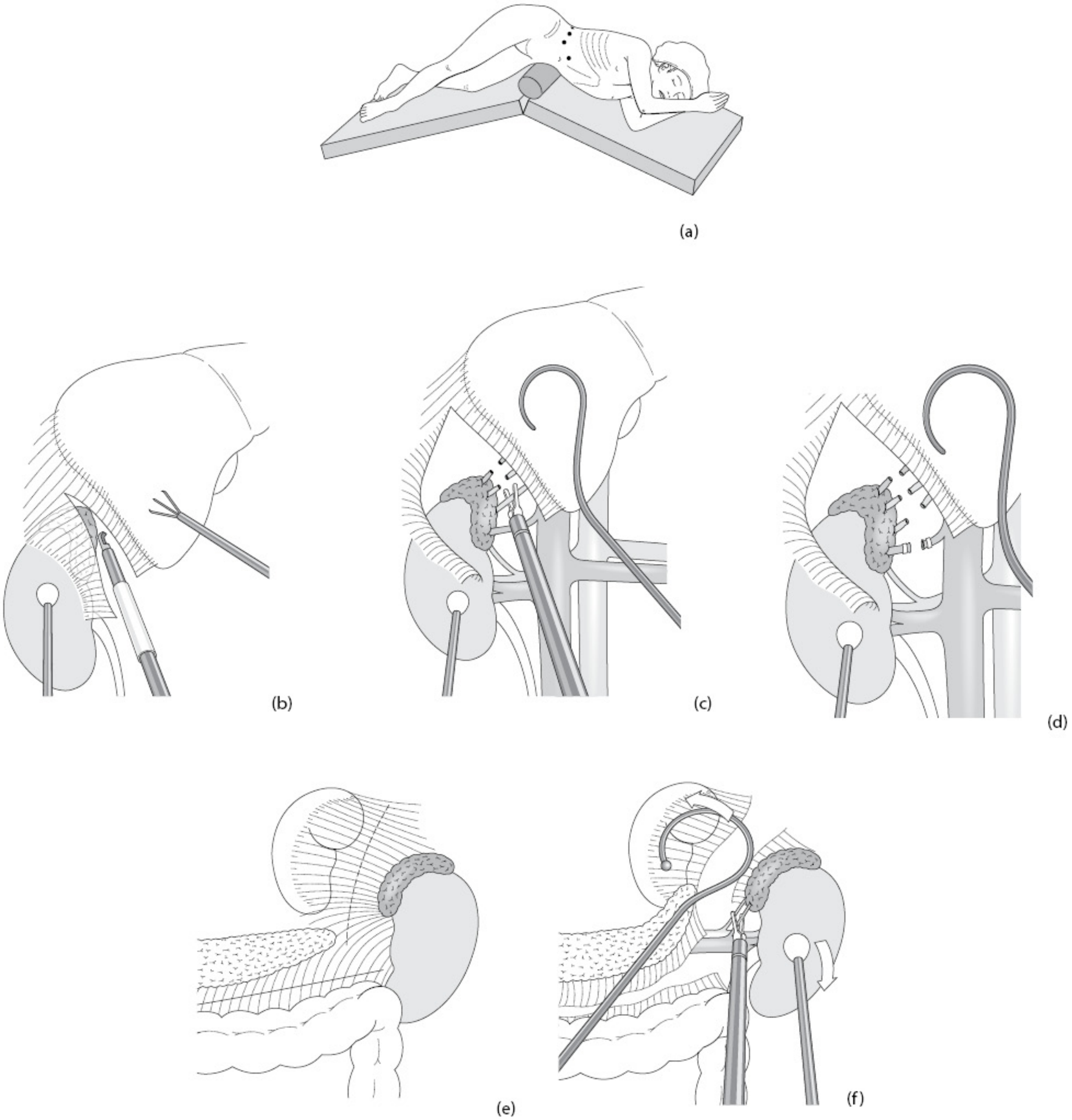
Laparoscopic adrenalectomy

Laparoscopic adrenalectomy has largely supplanted the open approach for resection of both benign and malignant lesions. Advantages include improved visualization with a magnified view of the operative field, less postoperative pain, and a shorter hospital course. Disadvantages include the need for repositioning for bilateral tumors and the requirement for advanced laparoscopic skills on behalf of the surgeon. While the exclusion criteria for laparoscopic adrenalectomy are ever diminishing, it is still relatively contraindicated for large malignant neoplasms or malignancies with potential lymph node involvement.

As in open procedures, both transperitoneal and retroperitoneal approaches for laparoscopic adrenalectomy have been reported. The transperitoneal approach is described here.

LAPAROSCOPIC RIGHT ADRENALECTOMY

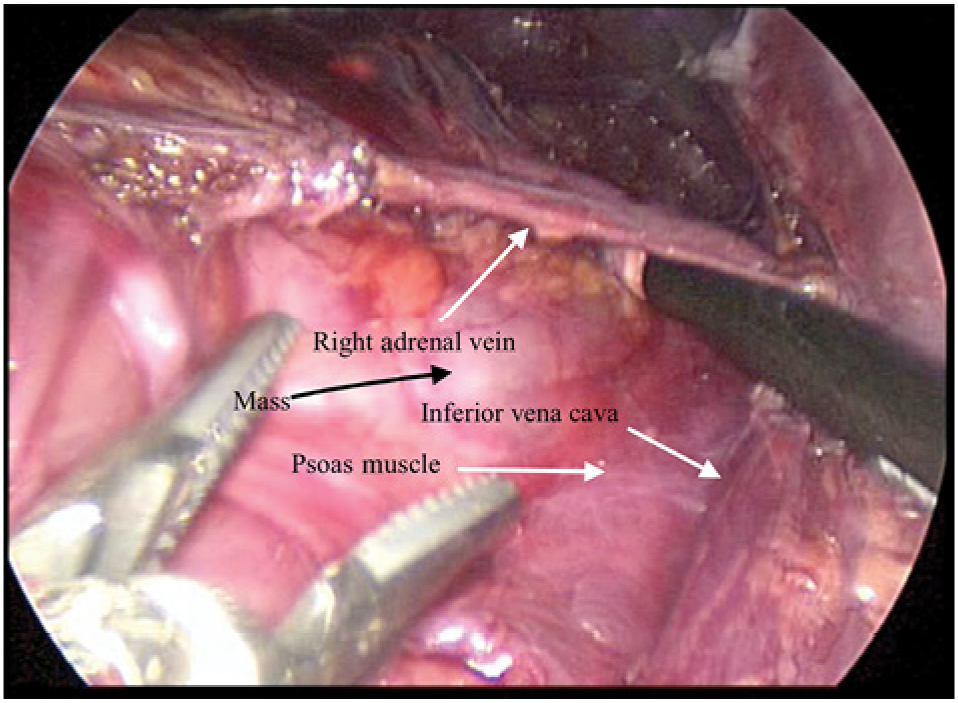
The patient is placed on a beanbag in the lateral decubitus position. Use of a kidney rest and table flexion at a point halfway between the ASIS and the costal margin opens the space between the iliac crest and the twelfth rib. An axillary roll is also put in place (**Figure 65.5a**). Either the true lateral decubitus or a gentle 15-degree rotation posteriorly off the true decubitus has been described. The ipsilateral arm should be supported on a mobile armrest. The patient may be secured in this position with tape. There is no need for a urinary catheter.



65.5a–f

The surgeon stands at the patient's abdomen opposite the surgical assistant. A second assistant (camera holder) may stand adjacent to the surgeon. Typically, four trocars are used for a transperitoneal approach. The position of the trocars may vary slightly depending upon patient body habitus. The first trocar (the optical trocar for a 5 mm, 30-degree scope or 10 mm, 30-degree scope) is inserted at the umbilicus. A second trocar is positioned approximately halfway between the umbilicus and the iliac crest. A third, operating trocar is placed about 5 cm anteromedial from the optical trocar, in the anterior axillary line. Another operating trocar is placed in the posterior axillary line at the same level. Occasionally, it is necessary to mobilize the right colon prior to placing this port. The fourth trocar is used for liver retraction and is placed medial and cephalad to the optical trocar. In larger patients, this may be placed 8–10 cm caudad to the xiphoid; in smaller patients, a supraumbilical site may be more useful. A triangular or curved liver retractor is placed through this port and secured to a self-retaining instrument holder.

Dissection begins through the lateral ports. Division of the right triangular ligament with L-hook electrocautery, Harmonic scalpel™ (Ethicon Inc., Cincinnati, OH, USA) (or LigaSure™ [Valleylab, Boulder, CO, USA] device) allows for better visualization of the adrenal (**Figure 65.5b**). The liver retractor is now positioned to allow the right lobe to be retracted superomedially. Division of the lateral duodenal attachments (Kocher maneuver) allows visualization of the IVC. The IVC is dissected superiorly and inferiorly, exposing the right renal vein (**Figure 65.5c and d**). The right renal vein marks the inferior point of caval dissection. The energy device is then used to dissect the peritonealized surface of the adrenal gland laterally, superiorly, and inferiorly (**Figure 65.5e**). As this dissection progresses, it should be possible to identify the adrenal vein, which may be visualized from its point of origin on the posterolateral wall of the vena cava (**Figure 65.6**). It should be mobilized for approximately 1 cm, doubly clipped, and divided. The lateral positioning of the patient facilitates this dissection.



65.6 Right adrenal vein and exposure as it enters the IVC.

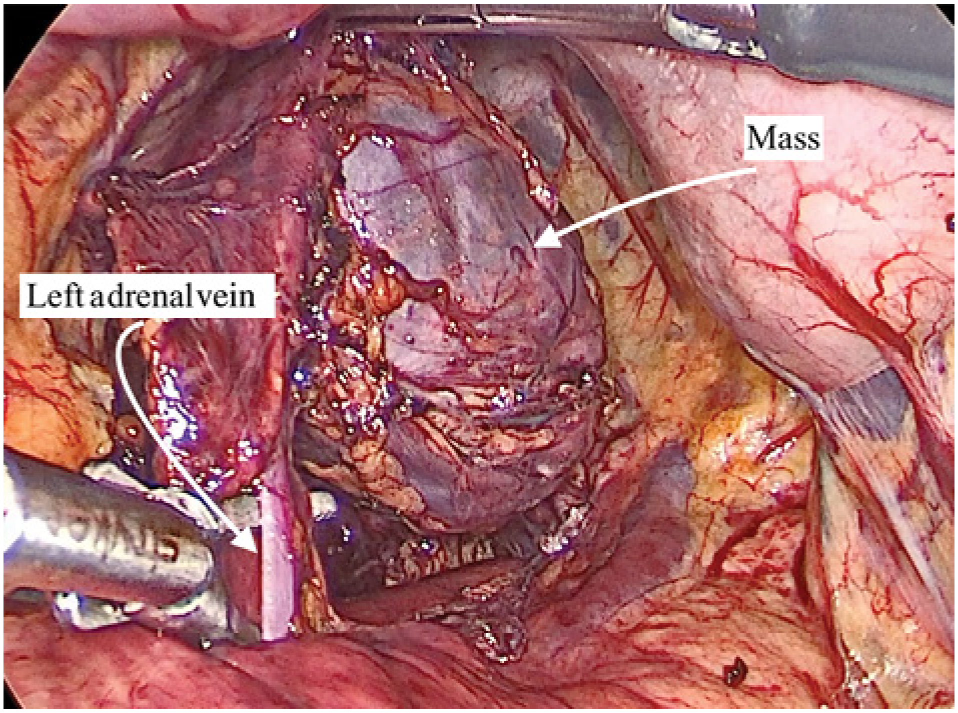
After the adrenal vein is divided, the adrenal should be gently retracted caudally. The gland itself is very friable and retraction should be done by gentle grasp of its peritonealized surface or by the use of laparoscopic Kitners. Occasionally (in 3–10% of cases), an accessory adrenal vein may also be visualized. After transection of the main adrenal vein, dissection should continue along the inferior aspect of the liver (superomedial aspect of the adrenal) to look for an accessory vein. This vessel should be dissected, clipped, and divided. The adrenal gland is then gently retracted caudally and to the right and dissected along its medial side. Arterial feeding vessels may be clipped and divided or cauterized. The laparoscopic energy devices are particularly good for this task. Once the upper pole of the adrenal is free, any remaining superior and lateral attachments of the adrenal may be dissected using hook electrocautery or an energy device (**Figure 65.5f**). The adjacent pole of the inferior medial attachments may contain the arterial supply from the renal artery. These attachments should be dissected carefully and any vessel of moderate size should be clipped prior to division or carefully cauterized using an energy device. At this point, the adrenal gland is attached only by loose posterior fibroareolar tissue. Once it is separated from this, it may be removed and placed into an extraction bag and removed from the abdomen. Typically, this will involve increasing one of the port sizes from 5 mm to 10 mm.

LAPAROSCOPIC LEFT ADRENALECTOMY

The patient is positioned as in a right-sided approach, except that the left side is elevated. It may be advantageous for the surgeon to work laterally to medially and thus stand at the patient's back, although an anterior position works well. Trocar positioning is similar to laparoscopic right adrenalectomy with the most medial port located along the linea alba and the most lateral port located at the mid to posterior axillary line.

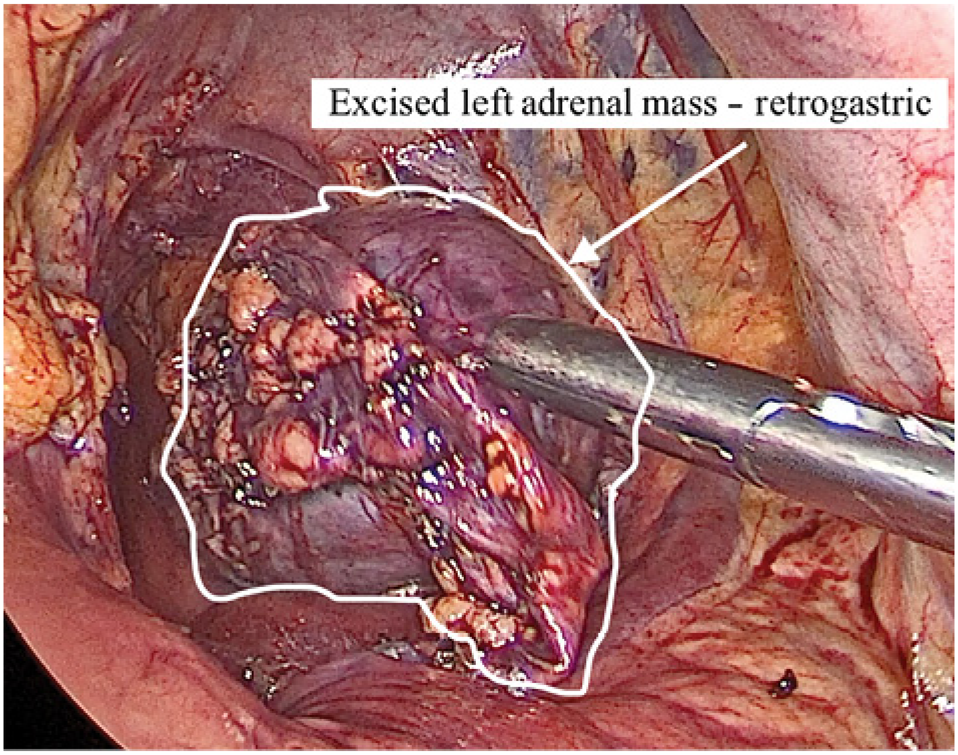
The dissection begins by reflection of the descending colon and splenic flexure down from the white line of Toldt and subsequent division of the lienophrenic and lienorenal ligaments. Dissection of the posterior spleen should be carried as far cephalad as the left diaphragmatic crus. The posterior surface of the pancreas may also be lifted off the splenic vein, permitting the spleen, pancreatic tail, and left colon to fall forward and medially. The retracting port may be used to assist in medial retraction of the spleen. Alternatively, the left adrenal gland may be approached from a retrogastric technique. The gastrocolic omentum is divided and the stomach is pexed to the anterior abdominal wall (see **Figure 65.3d**). Gerota's fascia should be opened vertically, beginning at the midpole of the kidney and moving superiorly. Superior to the upper border of the pancreas, a mass will be visualized. If the left adrenal vein has been clearly visualized draining into the left renal vein, it may be dissected, skeletonized, doubly clipped, and divided at this time.

Early division of the left adrenal vein is especially useful when operating on pheochromocytoma as it disrupts the release of catecholaminergic agents and facilitates better blood pressure control. If it is not visible, dissection along the superior aspect of the perinephric fat at the superomedial edge of the kidney should uncover yellow-orange tissue of the adrenal gland. The left adrenal gland may be dissected along its superior border. This dissection is greatly facilitated by the use of an energy device in which cauterization of small arterial branches from the left phrenic artery and aorta may be accomplished. Gentle anteroinferior retraction of the adrenal with a laparoscopic Kitner permits dissection of the lateral edge. If the left adrenal vein has not been yet identified and divided, this should now occur (**Figure 65.7**).



65.7 Revealing the left adrenal vein.

The final step in the dissection is to free the remaining tissue at the inferior and medial border of the left adrenal gland. Occasionally, two to three small arterial perforating vessels are present at this level, supplied by the superior branch of the renal artery, which should be divided. The free adrenal (**Figure 65.8**) may now be placed into an extraction bag and removed from the abdomen through an enlarged port site.



65.8 Left adrenal mass completely excised via retrogastric approach.

CONCLUSION

Many adrenal disorders require adrenalectomy. No one approach is superior to another, but the choice of resection must be tailored to the specific pathologic condition, patient anatomy, and surgeon experience. Laparoscopic adrenalectomy has been shown to be a safe and cost-effective approach to the resection of benign lesions. Its adoption has been widely broadened to include the resection of malignant lesions as well as small neuroblastomas, or the rare malignant pheochromocytoma.

VIDEOS

OPS_8e_video_65.1 Laparoscopic left adrenalectomy
OPS_8e_video_65.2 Laparoscopic right adrenalectomy

FURTHER READING

Guz BV, Straffon RA, Novick AC. Operative approaches to the adrenal gland. *Urol Clin North Am* 1989; 16: 527–34.

Mirallié E, Leclair MD, de Lagausie P, et al. Laparoscopic adrenalectomy in children. *Surg Endosc* 2001; 15: 156–60.

Rescorla FJ. Malignant adrenal tumors. *Semin Pediatr Surg* 2006; 15: 48–56.

Romano P, Avolio L, Martucciello G, et al. Adrenal masses in children: The role of minimally invasive surgery. *Surg Laparosc Endosc Percutan Tech* 2007; 17: 504–7.

Skarsgard ED, Albanese CT. The safety and efficacy of laparoscopic adrenalectomy in children. *Arch Surg* 2005; 140: 905–8.

St Peter SD, Valusek PA, Hill S, et al. Laparoscopic adrenalectomy in children: A multicenter experience. *J Laparoendosc Adv Surg Tech A* 2011; 21(7): 647–9.

Stewart JN, Flageole H, Kavan P. A surgical approach to adrenocortical tumors in children: The mainstay of treatment. *J Pediatr Surg* 2004; 39: 759–63.

PART VII

Urology and gynecology

66

Cystourethroscopy

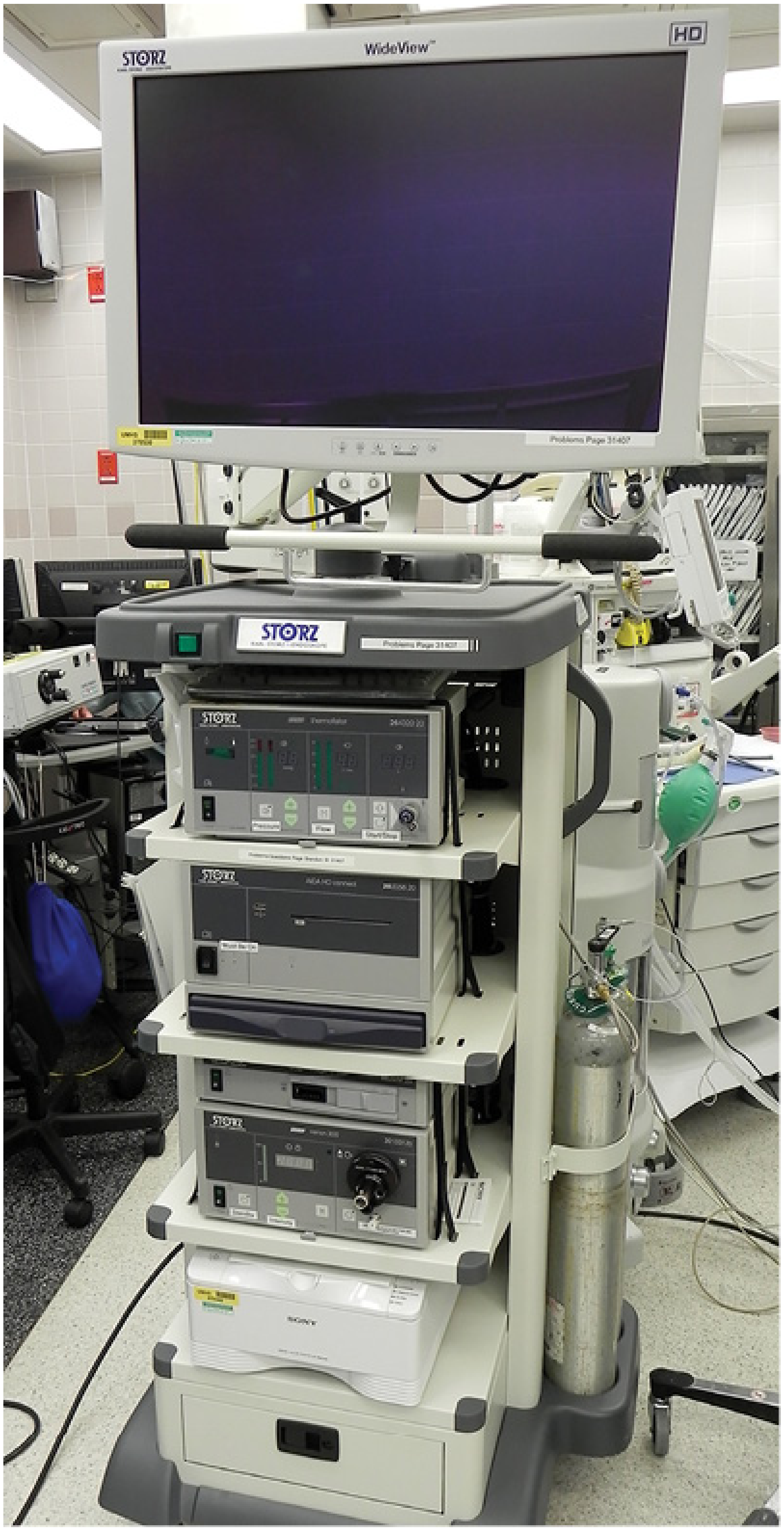
JOHN M. PARK

INTRODUCTION

In the surgical management of various congenital and acquired genitourinary problems, cystourethroscopy plays a critical role in both diagnosis and treatment. This chapter describes key principles of cystourethroscopy as it pertains to various urologic conditions including vesicoureteric reflux (VUR), ureteral duplication anomalies, ureterocele, posterior urethral valves (PUVs), genitourinary tumors, and urogenital sinus. It reviews the key principles of safe surgical practice of cystourethroscopy as well as various anatomic features of different conditions and their treatments.

SET-UP

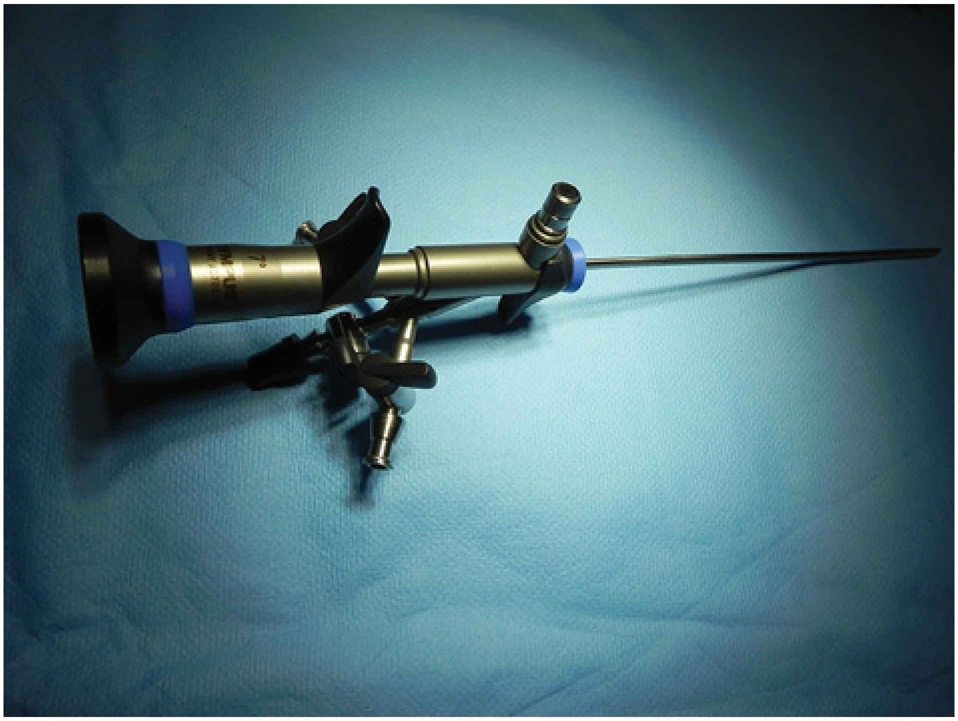
One advantage of modern endoscopic procedures is the ability to transmit a clear, magnified image onto a video system (**Figure 66.1**). A high-intensity light source, such as halogen and xenon, is necessary for proper illumination. Extreme care must be taken to avoid direct contact between the lighted cable and the patient or surgical drapes because of the intense heat generated. Hazardous fires and burn injuries can result. Continuous vigilance and communication among surgical team members is critical. The patient is placed into a dorsal lithotomy position. In infants, the legs may be simply "frog-legged" if extreme lateral scope deviation is not required.



66.1 Modern video tower fitted with monitor, light source, image processor, and recording device.

CYSTOURETHROSCOPE

The advancement of fiberoptic technology has led to the development of small caliber pediatric endoscopes. The outer sizes vary between 6 and 21 Fr (**Figure 66.2**). Infant resectoscopes have an outside diameter of 8.5 Fr. A variety of electrodes can be fitted for both cutting and coagulative procedures. The male neonatal urethra at full term can usually accommodate an 8.5 Fr instrument. The size is often dictated by the caliber of the urethral meatus. Gentle dilation or meatotomy may facilitate the insertion of scopes. The female urethra can, in general, accommodate between two and four Fr sizes larger than the male urethra.



66.2 A 7.9 Fr integrated fiberoptic infant cystoscope.

WORKING INSTRUMENTS

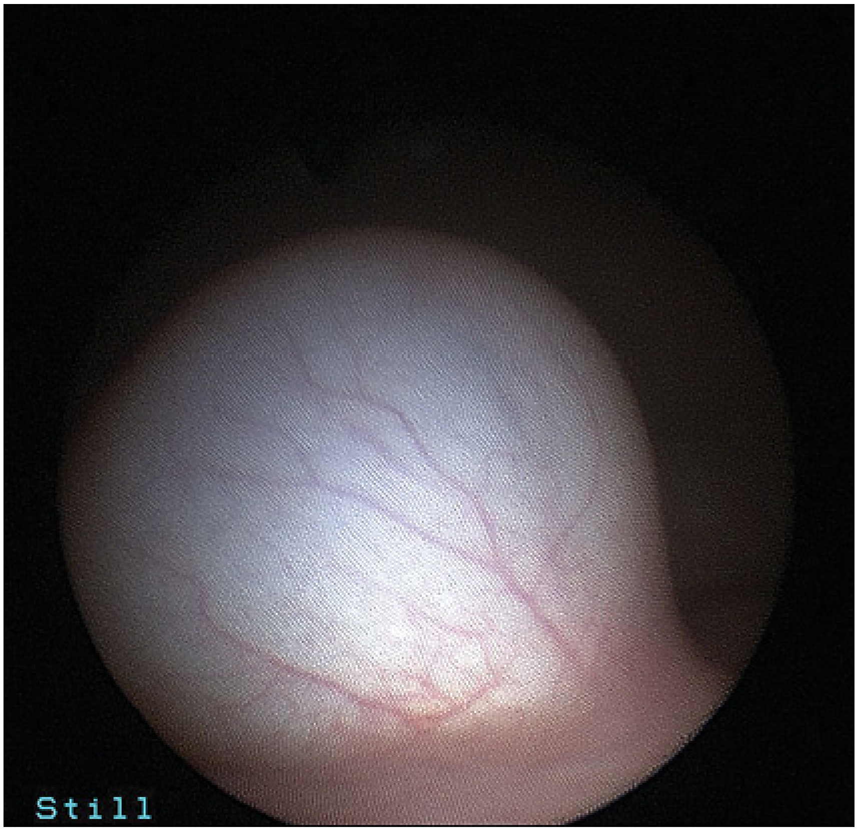
Guidewires are commonly used in endourology. Passage of catheters across ureteric orifices, urethral strictures, and false passages is accomplished over guidewires using the Seldinger technique. There are different diameters, ranging from 0.64 mm to 0.97 mm (0.018–0.038 inches). The distal tip may be straight or angled. The flexible tip varies in length from 1 cm to 15 cm. Some have a movable core wire, which offers varying lengths of distal flexibility. There are several types of coating, the most common being Teflon®. Hydrophilic polymer coatings (called "glidewires") decrease the friction of wires, making them useful for negotiating tortuous or narrow lumens. The slippery coating is activated by wetting with water or saline, and it should not be used with lubricant, which increases the friction.

Several cutting and fulgurating electrodes are available, in sizes 2–5 Fr. The electrical current is dispersed when using saline solution, and thus the irrigant should be either water or other osmotic solution, such as glycine, when using electrodes.

CLINICAL APPLICATIONS

Ureterocele

Ureterocele is a congenital obstructive cystic dilation of the submucosal portion of the distal ureter. An intravesical (or orthotopic) ureterocele is contained entirely within the bladder (**Figure 66.3**), whereas an ectopic ureterocele's distal portion is located within the bladder neck or the urethra. Ectopic ureteroceles represent approximately 60% of all ureteroceles, are more prevalent in females, and are commonly associated with ureteral duplication. In ureteral duplication, the ureteroceles belong to the upper- pole segment. The management of ureterocele depends on many factors, including the degree of associated ureteral dilatation and obstruction, the function of the associated renal moiety, and lower-pole ureter reflux. In many cases, endoscopic decompression using cystoscopy is an appropriate initial approach, especially in infants with urosepsis due to obstruction. Endoscopic decompression can result in secondary reflux, which may require subsequent intervention.

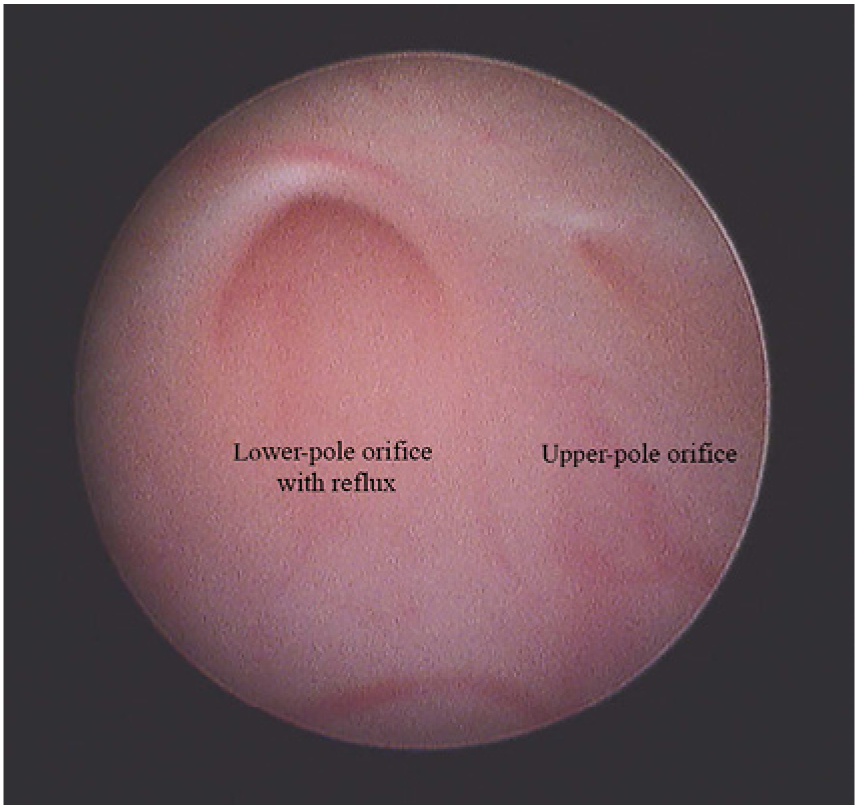


66.3 Intravesical ureterocele.

The visualization of a ureterocele is optimal when the bladder is not overly distended. With a full bladder, the ureterocele can efface and flatten, making it difficult to incise effectively. If there is a deficiency of bladder muscle backing at the site of the ureterocele, it will evert, making it look like a diverticulum. If the child is ill with urosepsis, a large transverse incision for definitive decompression is the primary goal, even if it results in secondary reflux. In the absence of clinical infection, a more limited incision or multiple punctures (like a “watering can”) can lead to effective decompression while minimizing the risk of secondary reflux.

Duplication anomalies

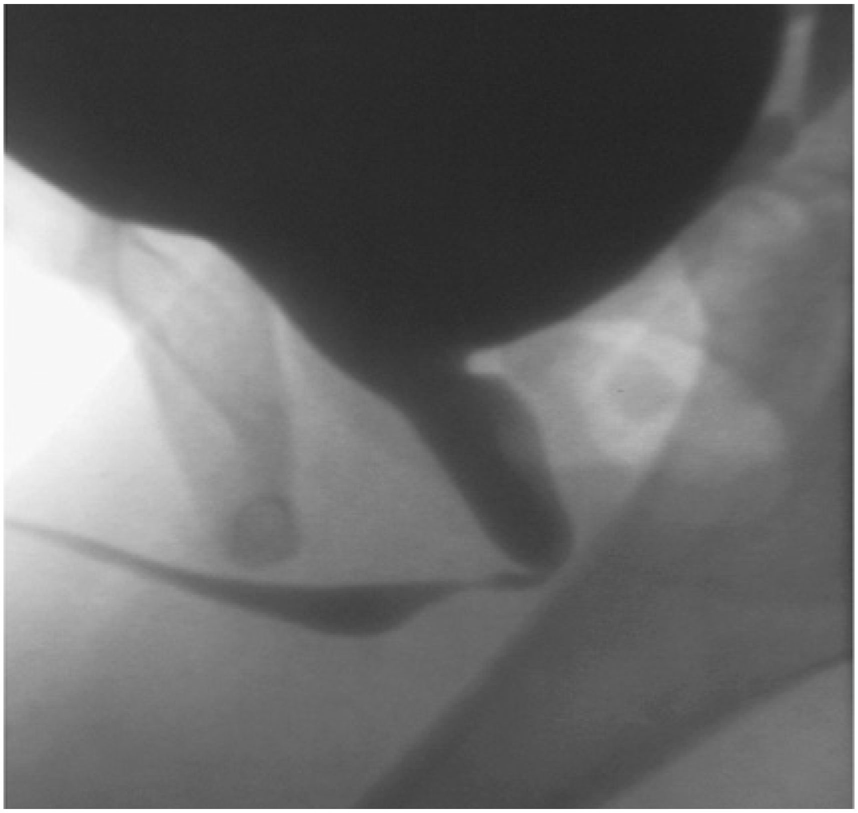
Although the true incidence of ureteral duplication is unknown, it is estimated to be approximately 1 in 125. According to the Weigert–Meyer rule of ureteral embryology, the two orifices of a complete duplication seen via cystoscopy are characteristically inverted in relation to the collecting systems they drain (Figure 66.4). The orifice to the lower-pole ureter occupies the more cranial and lateral position, and that of the upper-pole ureter has a caudal and medial position. In general, the lower-pole orifice tends to be associated with vesicoureteric reflux, while the upper-pole ureter tends to be ectopic, closer to the bladder neck and proximal urethra.



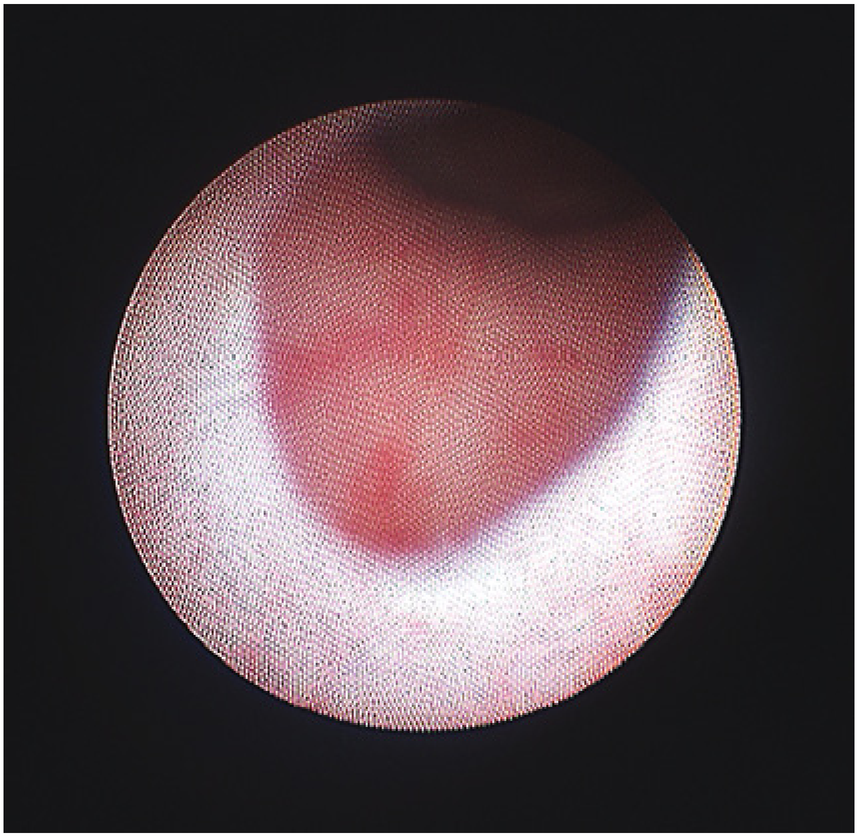
66.4 Duplicated orifices on the right with refluxing lower-pole orifice situated more laterally compared to more medially located upper-pole orifice.

Posterior urethral valves

PUVs are the most common congenital urethral obstruction in male newborns. It is often detected *in utero* based on prenatal sonography. Severe obstruction can lead to multiple perinatal issues due to renal failure and oligohydramnios, leading to respiratory and orthopedic problems. A voiding cystourethrogram (VCUG) will confirm the diagnosis, based on the appearance of a severely trabeculated bladder, thickened bladder neck, dilated posterior urethra, and an obstructive lumen change between the posterior and membranous urethra (**Figure 66.5**). Three types of valves have been described classically, although type II is no longer considered valid. Type I valves arise from the verumontanum with a "sailboat spinnaker" appearance to the leaflet (**Figure 66.6**). The visualization of type I valves is best accomplished by filling the bladder to its maximal capacity, positioning the scope near the distal portion of the posterior urethra at the verumontanum, and observing for the obstructing leaflets while the antegrade flow is induced with suprapubic pressure. Type III valves are located distal to the verumontanum and are diaphragmatic in appearance.



66.5 Voiding cystourethrogram demonstrating PUVs.

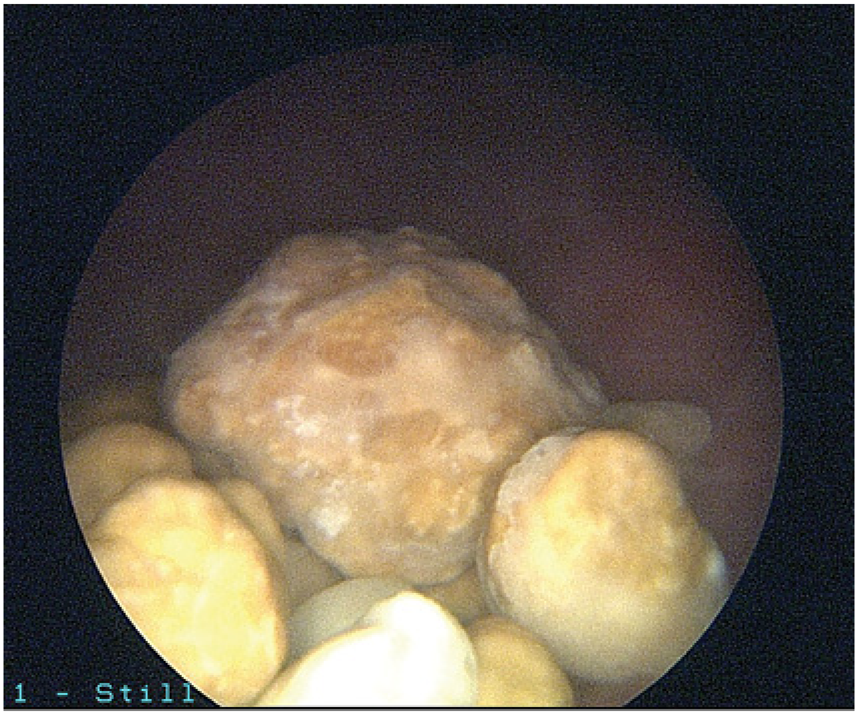


66.6 Obstructing leaflets of the PUVs.

Unless the newborn is extremely premature, the standard infant resectoscope with an 8.5 Fr sheath may be used for direct vision valve ablation. The hook blade is well suited for engaging the leaflet of the valves. An optical cold knife, electrosurgical incision, and laser ablation will all work well. The valve leaflets are ablated at the 5 o'clock and 7 o'clock positions. Sometimes there are residual obstructing valve remnants anteriorly, which may need additional ablation. For type III diaphragmatic valve obstruction, multiple stellate incisions are most effective. It is important to avoid injury to the external urethral sphincter located distal to the valves. Although the bladder neck appears obstructive, it should not be incised since it may cause excessive bleeding, urinary incontinence, and future retrograde ejaculation.

Bladder stones

The term "lithotomy" was coined in 276 BCE by the Greek surgeon Ammonious of Alexandria. The original Hippocratic oath used to contain a warning to leave the lithotomy procedure ("cutting for stone") to the specialists in this art, making the urologist the first surgical subspecialist. Because of diet poor in animal protein, endemic bladder stones were common until the time of the Industrial Revolution. Open cystolithotomy was the most common pediatric surgical procedure in the Great Ormond Street Hospital for Children in the 1900s. In modern pediatric urology and surgery practices, bladder stones occur in children with bladder outlet obstruction or neurogenic bladder dysfunction (**Figure 66.7**). In children undergoing enterocystoplasty for augmentation, the lifetime risk of bladder stone formation exceeds 30%. They are caused by urinary stasis, mucus production, and bacteriuria. Chronic bladder irritation by stone fragments can lead to future problems such as recurrent infections, hematuria, and malignancy. The removal of all stone fragments is essential to prevent a nidus for stone recurrence.

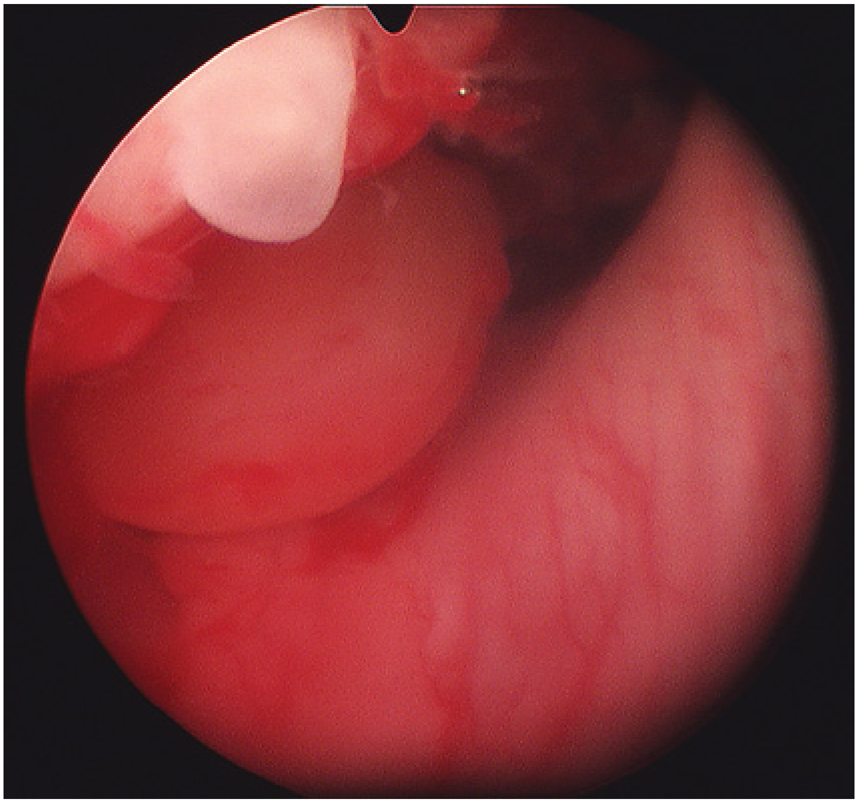


66.7 Bladder stones in the neurogenic bladder augmented previously with ileum.

The cystoscopic removal of bladder calculi is limited by the small caliber of the urethra in young children, especially boys. Various forms of intracorporeal lithotripsy have proven to be effective in fragmenting stones. The electrohydraulic lithotripsy (EHL) involves generation of an electric spark that produces a shock wave and a cavitation bubble for stone fragmentation. Other lithotripsy devices include ultrasonic, ballistic, and various forms of laser. It is important to take extreme caution and avoid direct contact with the bladder wall, as these techniques can lead to iatrogenic bladder perforation. For large or multiple bladder stones, retrieval of numerous stone fragments after lithotripsy can be quite difficult, and thus open cystolithotomy may be preferred. Percutaneous removal of bladder stones has also been described, especially in reconstructed bladders where the urethra has been closed.

Bladder tumor

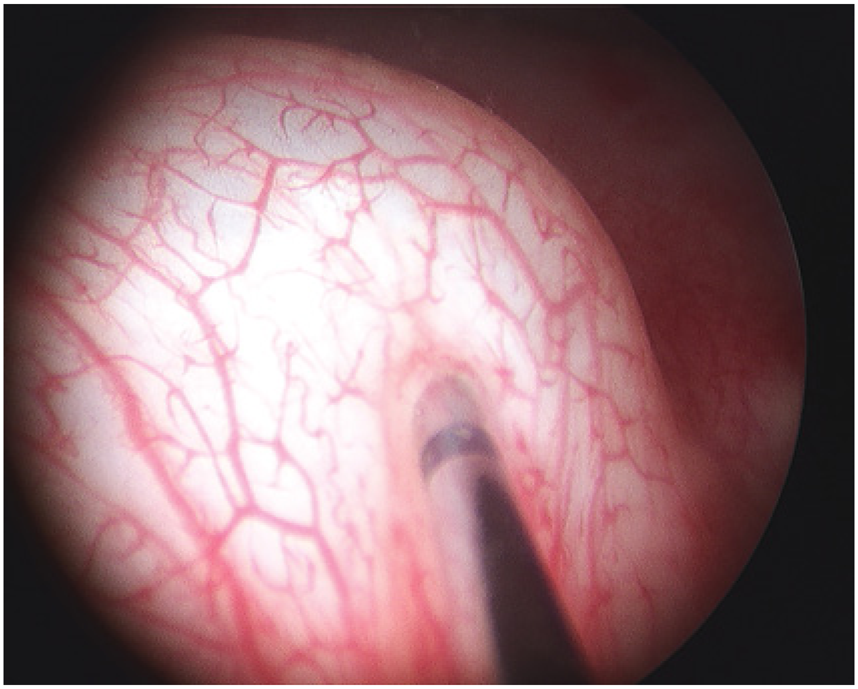
Approximately 15–20% of all pediatric rhabdomyosarcomas arise from the genitourinary system. The most common sites are prostate, bladder, and paratesticular. The rhabdomyosarcoma of bladder and prostate can present with urinary retention and hematuria. Imaging studies and cystoscopy will show a botryoid-type of sarcoma (**Figure 66.8**). In most cases, a cold-cup biopsy through the working channel of the cystoscope will yield the necessary tissue for diagnosis. If the child's urethra is large enough to accommodate the instrument, partial transurethral resection with a loop electrode will provide a generous tissue sample, deep enough for proper examination, as well as relieve obstruction.



66.8 Botryoid-type of bladder neck rhabdomyosarcoma.

Vesicoureteric reflux

VUR results from a combination of multiple factors, including the functional integrity of ureter, the anatomic status of the vesicoureteric junction (VUJ), and the bladder storage/emptying. In children without bladder outlet obstruction and neuropathic conditions (such as spina bifida and urethral valves), VUR is thought to represent a congenital defect of the VUJ. The abnormal ureter length to diameter ratio of the intramural ureter leads to a failure of the "flap-valve" mechanism. VUR diagnosis is made by VCUG, which provides a reproducible classification of grading, as well as other pertinent anatomic details that can help in the management decisions. The cystoscopic evaluation of children with VUR is no longer considered a routine part of the diagnostic workup. In the past, the characterization of the endoscopic appearance of the ureteric orifices was incorrectly thought to help in assessing the prognosis. In the modern VUR management, cystoscopy plays a major therapeutic role via subureteric injection. Various materials have been injected, including polytetrafluoroethylene paste, polymimethylsiloxane, and bovine collagen, with varying degrees of success and risk. Currently, the dextranomer–hyaluronic acid copolymer (Deflux®) is the only agent approved by the US Food and Drug Administration for endoscopic VUR treatment. The success rate varies in direct proportion to the grade of reflux. Some have reported a short-term success rate of 90% in low-grade VUR after a single Deflux injection. The ureteric orifice is hydrodistended with irrigant aimed at it. The orifice associated with high-grade reflux tends to distend widely and, in these situations, it is recommended to position the needle and inject into the floor of the intramural ureter, followed by a classic subureteric injection to elevate the lower lip of the orifice, causing a "volcano"-shaped opening (**Figure 66.9**).



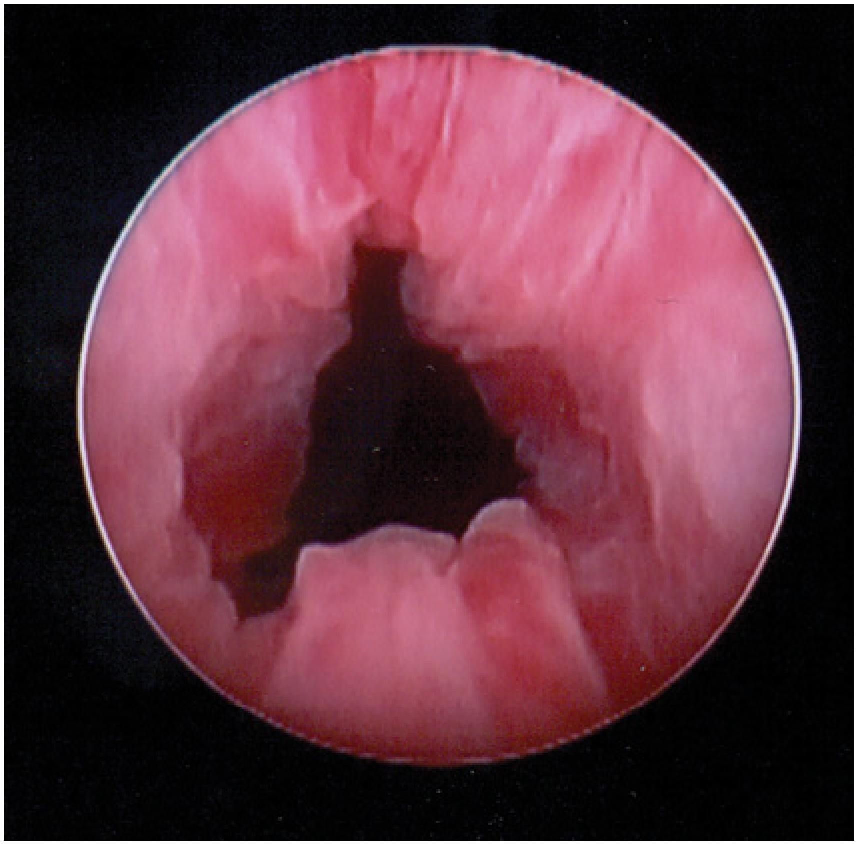
66.9 Deflux injection.

Incontinence

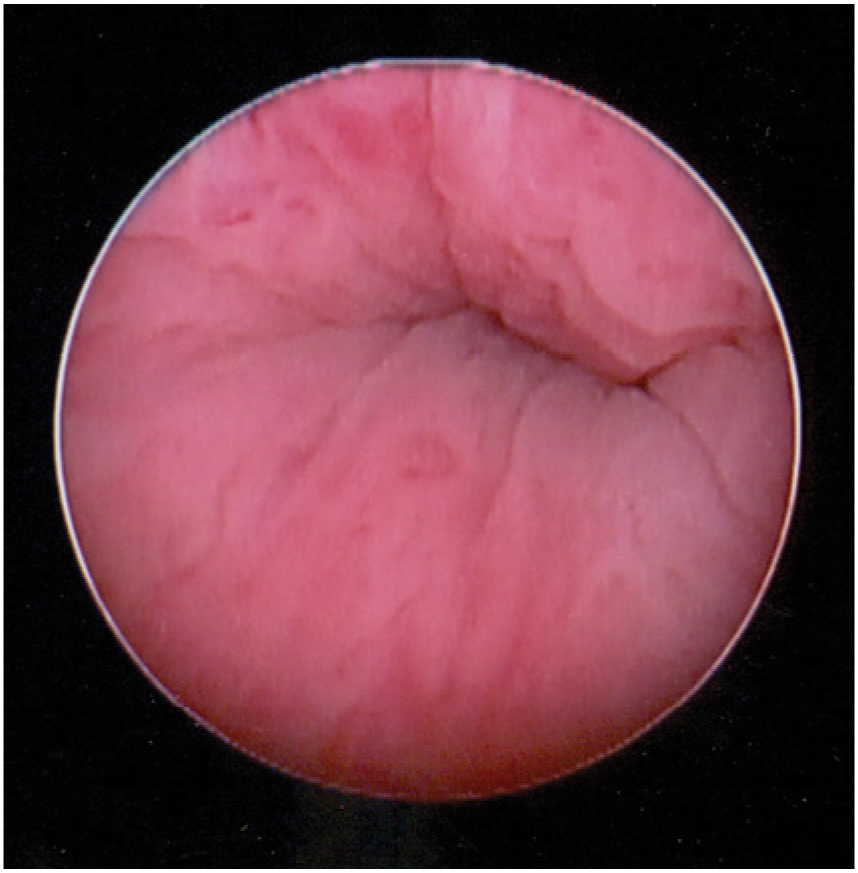
Incontinence secondary to anatomic abnormalities, such as congenital neuropathic conditions (spina bifida, sacral agenesis), exstrophy–epispadias complex, and bilateral ectopic ureters, requires intervention. Endoscopic implantation of bulking agents is a simple and logical attempt to increase outlet resistance with minimal morbidity. The treatment strategy for incontinence must be developed within the physiological framework of normal bladder function. The hallmarks of a normal lower urinary tract include safe storage pressures, ample capacity, effective outlet resistance that can withstand intra-abdominal pressures, and, finally, complete periodic emptying of urine. Incontinence is rarely an isolated matter of inadequate outlet resistance alone. Most of the endoscopic treatment strategies for incontinence have focused on improving the quality of bladder outlet closure mechanism by implanting various bulking agents.

Injection of a bulking agent into the submucosal space of the urethra and bladder neck can be accomplished by two approaches.

- In the *transurethral approach*, the injection needle is passed through the working channel of the cystoscope, and the needle is positioned into the submucosa of the bladder neck and proximal urethra under direct vision. The substance is injected slowly, while observing the bulging of the mucosa into the lumen (**Figures 66.10 and 66.11**). In females, the bulking agent may be placed either transurethrally or periurethrally.



66.10 Open bladder neck causing incontinence.



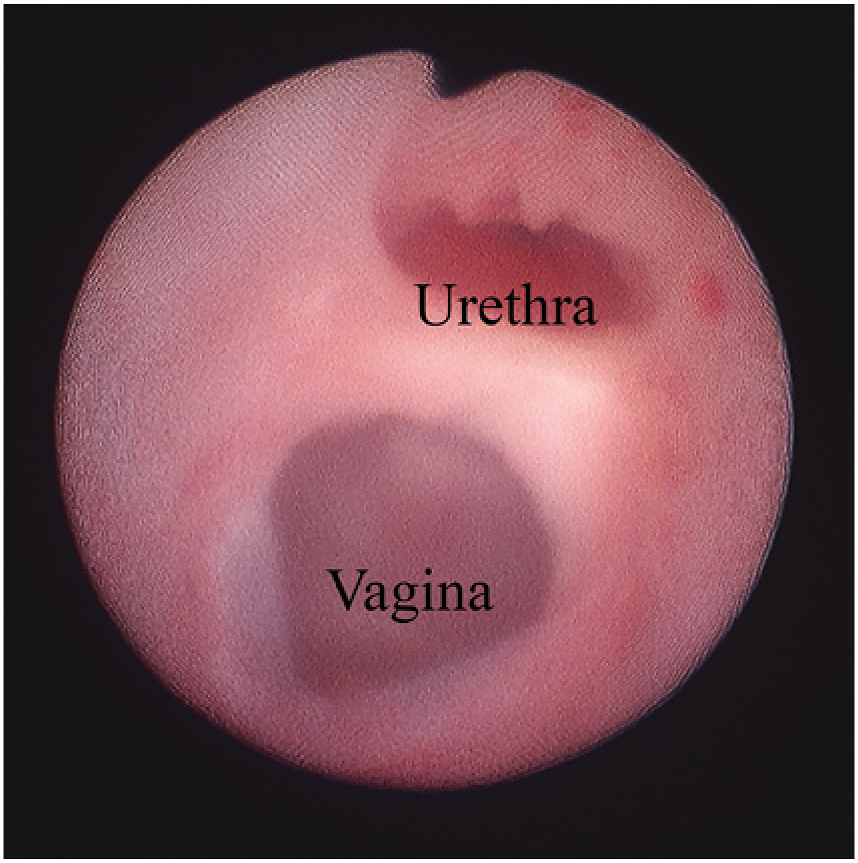
66.11 Bulging mucosa at the bladder neck after Deflux injection.

- In the *periurethral approach*, the injection needle (typically, a long 25 gauge) is passed through the anterior vaginal wall to the appropriate submucosal location at the bladder neck, and the injection is performed under direct cystoscopic vision. The general principle is that the closer the injection occurs to the bladder neck and proximal urethra, the more effective the outlet mechanism becomes against stress urinary incontinence.

Recently, endoscopic treatment has been used in select patients in whom the primary abnormality is one of detrusor dysfunction of uninhibited contractions (bladder spasms) and stiffness (poor compliance). In these situations, therapy is aimed at modification of detrusor function to improve bladder storage and emptying. Onobotulinum toxin A induces a muscle paralysis by blocking the fusion of acetylcholine-containing vesicles with the cell membrane, thereby impeding the release of acetylcholine into the synaptic space. The most common urologic indication has been the injection of onobotulinum toxin into the external urethral sphincter for the treatment of detrusor–sphincter dyssynergia, primarily in patients with spinal cord injury. Onobotulinum toxin may also be injected directly into the detrusor muscle to decrease contractility and increase functional capacity.

Urogenital sinus abnormalities and cloacal malformation

Urogenital sinus (UGS) abnormalities are most often seen in intersex cases, now referred as "disorders of sex development" (DSD). In repairing the UGS abnormalities, certain critical details of the anatomy, including the length of the common UGS channel, the location of the vaginal confluence, its proximity to the bladder neck, the size/number of vaginas, and the presence of a cervix, must be defined (**Figure 66.12**). In patients with congenital adrenal hyperplasia (CAH), endoscopy is usually performed at the time of reconstruction. Some of these female CAH patients will demonstrate a male-like external urethral sphincter with the vagina entering proximal to it in a verumontanum-like area. As with UGS abnormalities, endoscopy is mandatory to define the anatomy of a cloacal malformation. The goals of endoscopy must include identification of the rectal and vaginal confluence. The length of the urethra and its communication with the cloaca are important for reconstructive purposes. Vaginal duplication is frequently seen. The rectal confluence is most commonly located at the level of vaginal confluence.



66.12 Vaginal confluence in a congenital adrenal hyperplasia patient with urogenital sinus.

FURTHER READING

Chertin B, deCaluwe D, Puri P. Is primary endoscopic puncture of ureterocele a long-term effective procedure? *J Pediatr Surg* 2003; 38: 116–19.

Dinneen MD, Duffy PG. Posterior urethral valves. *Br J Urol* 1996; 78: 275–81.

Kirsch AJ, Perez-Brayfield MR, Scherz HC. Minimally invasive treatment of vesicoureteral reflux with endoscopic injection of dextranomer/hyaluronic acid copolymer: The Children's Hospitals of Atlanta experience. *J Urol* 2003; 170: 211–15.

Pena A, Levitt MA, Hong A, Midulla P. Surgical management of cloacal malformation: A review of 339 patients. *J Pediatr Surg* 2004; 39: 470–9.

67

Ureteropelvic junction obstruction

LAUREN E. CORONA AND KATE H. KRAFT

PRINCIPLES AND JUSTIFICATION

The ureteropelvic junction (UPJ) is the most common site of obstruction in the pediatric upper urinary tract. The UPJ is formed during the fifth week of embryogenesis. Urine flows across it from the kidney and down the length of the ureter by the twelfth week. Flow occurs when renal pelvic pressure exceeds upper ureteral pressure. The pressure gradient is created partly by the hydrostatic force of the filtered urine but principally by the peristaltic contractions originating in the region of the renal pelvis called the pacemaker. Hydronephrosis develops either when the UPJ is obstructed or when the normal peristaltic waves are impeded.

Obstruction most commonly results from an intrinsic defect in the smooth muscle layer of the UPJ. External compression and blockage by aberrant renal vessels and adhesive bands occur in approximately one-third of cases, and often a combination of external compression and intrinsic pathology exists. Light microscopy demonstrates that abnormal smooth muscle architecture and increased fibrosis are present in obstructed UPJs. Electron microscopy further shows disruption of the intercellular junctions needed to coordinate the transmission of peristaltic waves.

Incidence, signs, and symptoms

UPJ obstruction affects approximately 1 : 1500 live births and has a ratio of 2 : 1 in boys compared to girls. The left side is affected in approximately 60% of cases, and 5–10% of cases are bilateral. In duplicated systems, the lower-pole moiety is more likely to be obstructed, although both systems can be involved.

There are two groups of patients that present with UPJ obstruction. Those that present in the older age group will usually require surgical intervention, whereas the prenatally diagnosed hydronephrosis, which constitutes the larger proportion of patients, may not require surgery. The challenge for pediatric urologists is to identify those that warrant early surgical intervention against the majority of congenital hydronephrosis that resolves and can be safely observed.

Before the use of antenatal ultrasonography (US) became widespread in the 1970s, the common signs and symptoms of UPJ obstruction were abdominal mass, gross hematuria, urinary tract infection, and pain. Antenatal US now allows UPJ obstruction to be identified before becoming symptomatic and has increased the detection of asymptomatic obstruction fivefold, whereas the incidence of symptomatic obstruction has remained the same.

Older children and adolescents may have protean manifestations, including failure to thrive, vague sporadic flank and abdominal pain (especially with diuresis), nausea, vomiting, hypertension, and recurrent urinary tract infections. Hematuria or renal parenchymal injury may ensue from only minor trauma. Stasis of urine due to obstruction can increase the risk of kidney stone formation.

Particular attention should be paid to infants with other congenital anomalies. Conditions associated with UPJ obstruction include imperforate anus, congenital heart disease, esophageal atresia, and the VACTERL association (vertebral defects, imperforate anus, cardiac anomalies, tracheoesophageal fistula, radial anomalies, and renal dysplasia), as well as other urologic anomalies including contralateral multicystic dysplastic kidney, vesicoureteric reflux, and renal ectopia including horseshoe kidney.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

Imaging studies

Clinically suspected UPJ obstruction should be confirmed by imaging studies. US is non-invasive, radiation-free, and well tolerated. Characteristically, the UPJ produces a central lucency, which is the renal pelvis surrounded by communicating dilated calyces. A voiding cystourethrogram is advisable for all patients suspected of obstruction to rule out vesicoureteric reflux and other lower tract causes of hydronephrosis, such as posterior urethral valves. Although less commonly performed, IV pyelography can further outline upper tract anatomy, and delayed films will best visualize the UPJ. The low glomerular filtration rate of neonates precludes the use of IV pyelography.

Diuretic renal scintigraphy is particularly helpful in diagnosing and quantifying the severity of obstruction. The most common radionuclide is MAG3 (^{99m}Tc-mercaptoacetyltriglycine). A urethral catheter should be inserted in all patients undergoing a renal scan when evaluating obstruction. It is best to wait a few weeks until the newborn kidney has developed the ability to concentrate urine at around 44 weeks' gestation, as early functional evaluation may lead to misleading results on the MAG3 scan. A full bladder can exert back-pressure on the ureters and obscure the results. The scan produces two important results:

- It determines the percentage of total renal effort contributed by each kidney. A poor or non-functional kidney should be considered for removal rather than repair.
- The time it takes for half the radionuclide to wash out of the kidneys after administration of diuretic ($t_{½}$) is calculated. A $t_{½}$ of more than 20 minutes is most commonly associated with obstructed systems. Recent studies in infants with antenatally detected hydronephrosis suggest that an elevated half-time alone may not always be associated with clinically significant UPJ obstruction.

Advances in digital imaging algorithms allow computed tomography (CT) and magnetic resonance imaging (MRI) to produce excellent images in the sagittal and coronal planes in addition to the traditional transverse images.

These allow excellent visualization of renal anomalies, such as horseshoe kidney, although their cost limits their widespread use. CT involves sizable doses of radiation and, for children, magnetic resonance urography (MRU) is an attractive alternative. Dynamic gadolinium control enhanced MRU has been shown to be promising. It offers the same data of renal scintigraphy (percent function and $t_{1/2}$), as well as a clear depiction of the kidney, pelvis, and ureter. MRU is more expensive and requires young patients to be anesthetized to avoid motion artifact.

Alternatives to pyeloplasty

The aims of surgery are to improve drainage of the UPJ, thereby alleviating pain and preventing urinary tract infections (UTIs), kidney stones, and renal deterioration. For these reasons, significant obstruction is best repaired promptly.

The management of UPJ obstruction detected by antenatal US but remaining asymptomatic in the infant and young child has evolved over the past several decades. Many newborn kidneys continue to grow and develop normally despite imaging studies consistent with mild UPJ obstruction.

Endourological techniques used for the treatment of ureteral calculi have been adapted to the treatment of UPJ obstruction. The obstructed segment is incised and dilated through either a percutaneous or a retrograde approach and is left to heal over an indwelling stent for several weeks. Reports suggest that this method may be useful in older children after previous failed pyeloplasty, but we believe it has proven to be unsuitable for infants and children perhaps due to the smaller diameter of the ureter and the relatively larger size of the instruments currently available. Other constraints include large redundant pelves and crossing vessels.

OPEN PYELOPLASTY

The technique described is a dismembered pyeloplasty (Anderson–Hynes procedure). Unlike methods using flaps and advancements, it excises the pathologic segment and may be applied to a variety of anatomic configurations.

Anesthesia

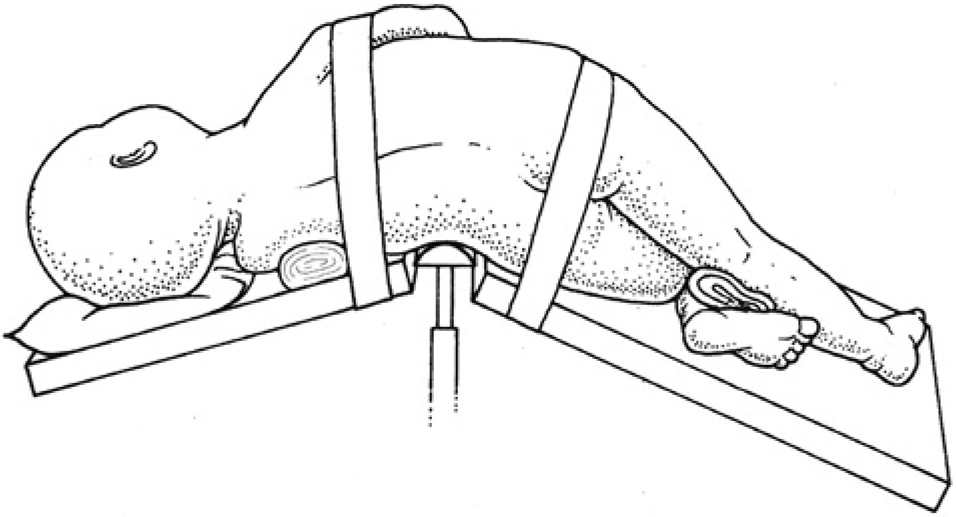
General anesthesia with endotracheal intubation is required and spinal morphine injection or other forms of regional anesthesia are helpful in reducing the total amount of general anesthesia used and providing pain relief postoperatively. Cystoscopy and intraoperative retrograde pyelography ensure there are no other regions of ureteral narrowing. This step can be skipped if a previous imaging study has cleared the length of the ureter.

Instruments and equipment

In addition to a pediatric major surgery tray, fine forceps, magnifying loupes, 3.5 Fr and 5 Fr feeding tubes, fine dissection scissors, needlepoint electrocautery, and 3/0 to 7/0 synthetic or biologic absorbable suture with atraumatic needles should be available. While the choice of suture varies greatly among surgeons, permanent synthetic or biologic sutures (e.g. silk or Ethibond™ polyester suture; Ethicon Inc., Cincinnati, OH, USA) should not be used because they can become nidi of infection and stone formation.

Position of patient

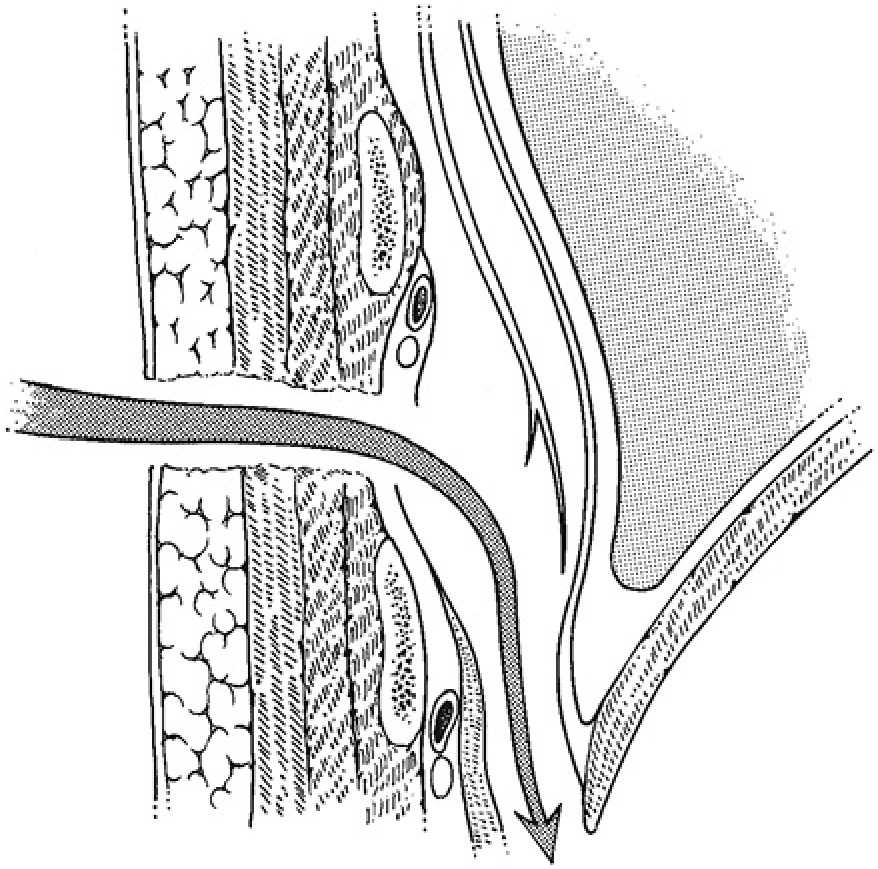
After intubation, a Foley catheter is placed and the patient is positioned. With the patient in the lateral decubitus position, the surgeon should stand facing the patient's back, with the assistant opposite. Padding and support should be placed under all pressure points. A kidney roll and axillary roll are also placed. The kidney rest is elevated and the table flexed. In small children, a modified anterior approach can be employed, with a small roll under the ipsilateral shoulder and thoracic side. The patient should be secured to the table with tape to permit lateral rotation of the table and enhance visualization (Figure 67.1).



67.1

Incision

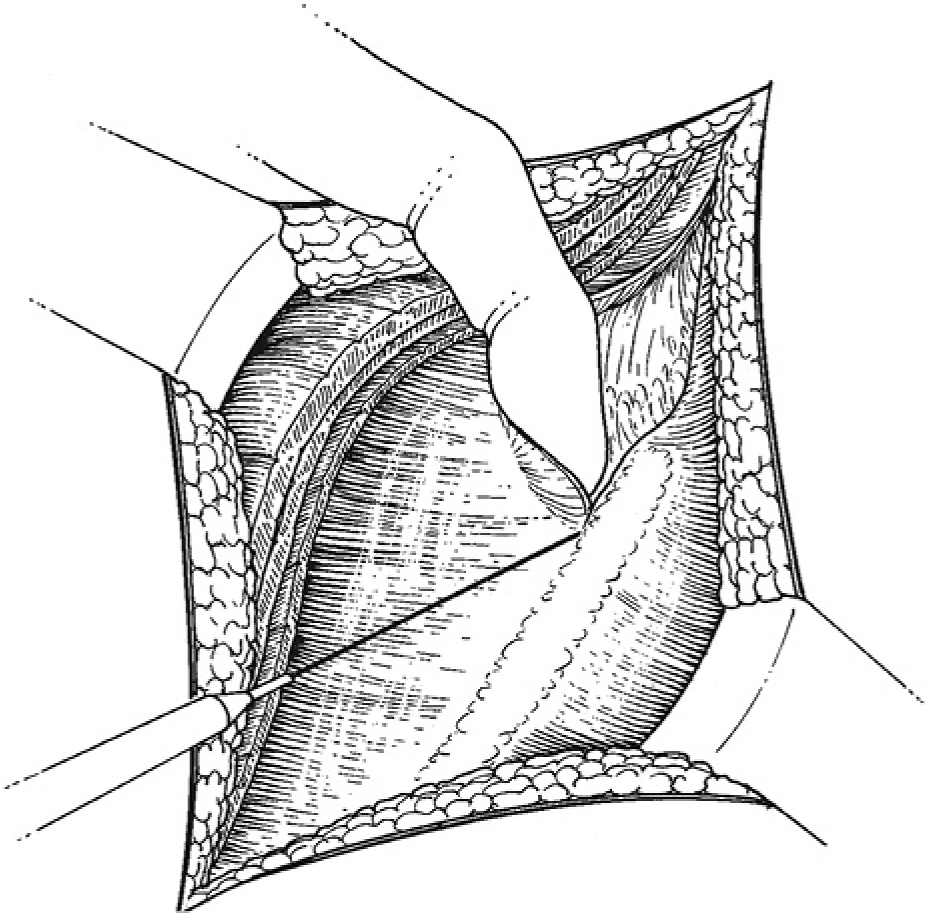
A supracostal, extrapleural flank approach is preferred. The skin incision is made astride the distal twelfth rib and extended anteriorly following the natural skin creases toward but short of the umbilicus. The external oblique and internal oblique, the latissimus dorsi, and the anterior edge of the serratus posterior muscles are divided until the periosteum of the rib is exposed (**Figure 67.2**). Other approaches include dorsal lumbotomy, anterolateral muscle splitting, and anterior transperitoneal. Lumbotomy allows direct access to the renal pelvis but creates a scar that crosses normal skin folds, and there is limited mobility of the kidney should further maneuvers be necessary, such as bringing the UPJ anterior to a crossing vessel. The muscle-splitting approach is suitable in infants and young children, with a modified anterior position. A supine transperitoneal approach is useful when access to the abdominal contents or the contralateral kidney is required. It is also useful when the UPJ obstruction is only one part of a larger urinary tract reconstruction (e.g. augmentation cystoplasty and UPJ obstruction repair). When performing a reoperative pyeloplasty, consideration should be given to either an alternative approach or extending the incision anteriorly beyond the current scar to allow entry into a pristine area and facilitate subsequent dissection.



67.2

Releasing the diaphragm

Entry into the retroperitoneum is made at the rib tip and extended posteriorly along the superior edge of the rib (**Figure 67.3**). The intercostal muscles should be carefully taken off the rib, keeping above the rib to avoid the neurovascular bundle. The diaphragmatic fibers are freed from their attachment to the rib using sharp dissection. The translucent pleura falls away as the diaphragmatic fibers are released. Patience and gentle dissection allow the fibers to move away naturally. The peritoneum is carefully peeled away from Gerota's fascia and retracted medially to allow for more working room.



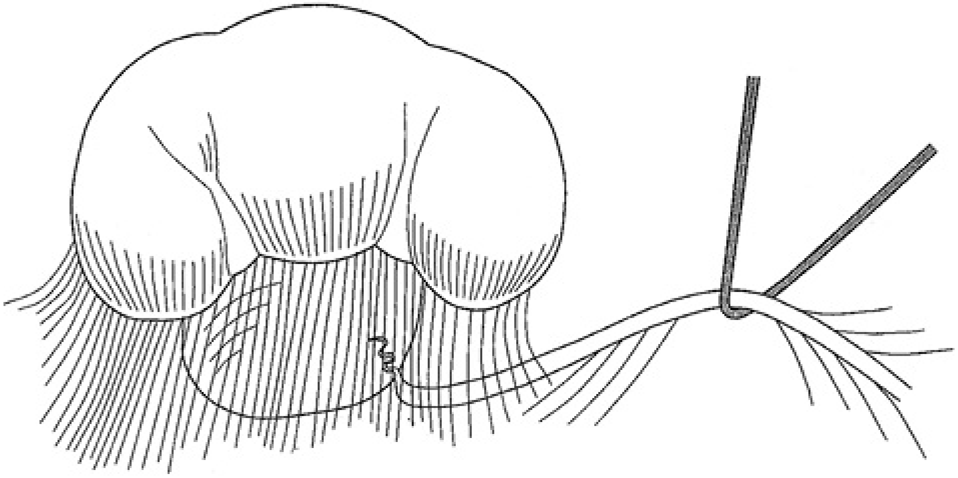
67.3

Kidney mobilization

A self-retaining retractor may aid exposure. Gerota's fascia is opened longitudinally and the overlying fat dissected away, taking care not to strip the renal capsule, which may be adherent. Patients who may have had renal leak or a decompressive nephrostomy tube may also have an inflamed, scarred layer of fat. If possible, the dissected fat as a single layer should be maintained, as it will be useful at the end of the case. Small blood vessels are coagulated. Branches and tributaries of the renal vessels around the anterior hilum should be sought. It is rarely necessary to skeletonize the vessels. The kidney should be mobilized sufficiently to expose the renal pelvis. The kidney can be held either by a well-padded retractor or by an assistant's hand. Displacing the kidney out of the depths of the incision may improve exposure, but exaggerated positions should be avoided because of the stretch imposed on the renal vessels and the risk of thrombosis or avulsion. In reoperative cases, approaching along the posterior aspect may be the safest way to begin mobilization; the hilum will be directly anterior with no major vessels to encounter before the pelvis.

Identifying the ureter

The ureter is identified and traced toward the kidney. Care should be taken to avoid stripping away the supporting adventitial vessels. One should be cautious on the left side not to confuse the gonadal vessels, which can have a similar course. Fine stay sutures and vessel loops are used to manipulate the ureter gently (**Figure 67.4**). The authors avoid grasping the pediatric ureter with forceps and prefer a "no touch" technique when possible. Aberrant vessels, particularly those near the lower pole of the kidney, should be identified and carefully dissected away but not divided.



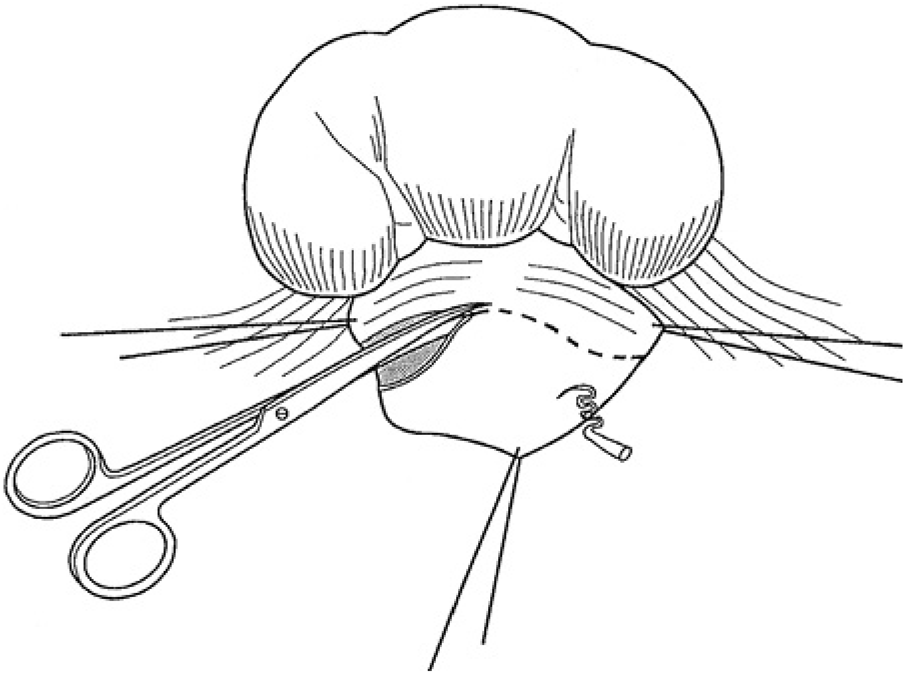
67.4

Preparing the renal pelvis and dividing the UPJ

The UPJ is identified and the overlying fat and other attachments cleared away. The type of UPJ encountered (e.g. high insertion), presence of crossing vessels, or other findings causally related to the obstruction should be carefully observed and documented. The ureter is divided just below the UPJ and controlled by a stay suture. If cystoscopy and retrograde pyelography were not performed prior to surgery, the ureter should be intubated with a small feeding tube or access catheter to ensure no distal narrowing. The ureter should be gently moved to check that adequate length has been mobilized. If insufficient length is available, further mobilization is necessary. Stay sutures are next placed on the renal pelvis at points lateral, medial, inferior, and superior to the UPJ to define the extent of the pyeloplasty.

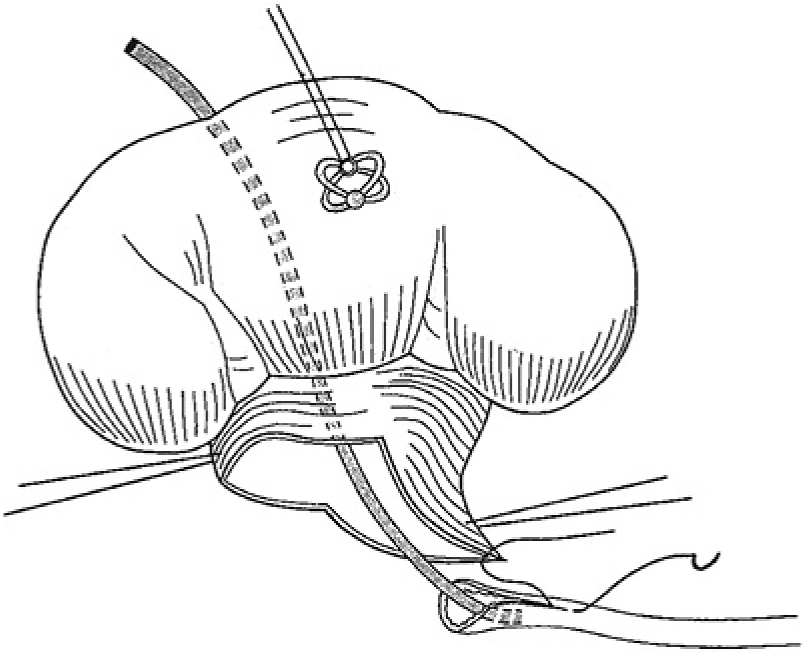
Excision of the UPJ and spatulation of the ureter

Traction is applied to the stay sutures to splay the decompressed renal pelvis flat. One smooth cut is made using sharp tissue scissors; repeated cuts tend to result in saw-tooth, jagged edges (**Figure 67.5**). A sharp hooked or pointed scalpel may be used instead of scissors. Irregular tags should be carefully trimmed. It should be borne in mind that if the patient has had a history of pyelonephritis or an indwelling nephrostomy catheter or stent, the wall of the pelvis might be thick. Additional pelvis may be excised if needed to reconfigure it easily into a funnel-shaped pyeloplasty. Debris or calculi are irrigated from the kidney. The course of the ureter is studied and noted. It is spatulated along the lateral aspect to meet with the lower extent of the pelvis without resulting in a twist or torsion of the ureter.



67.5

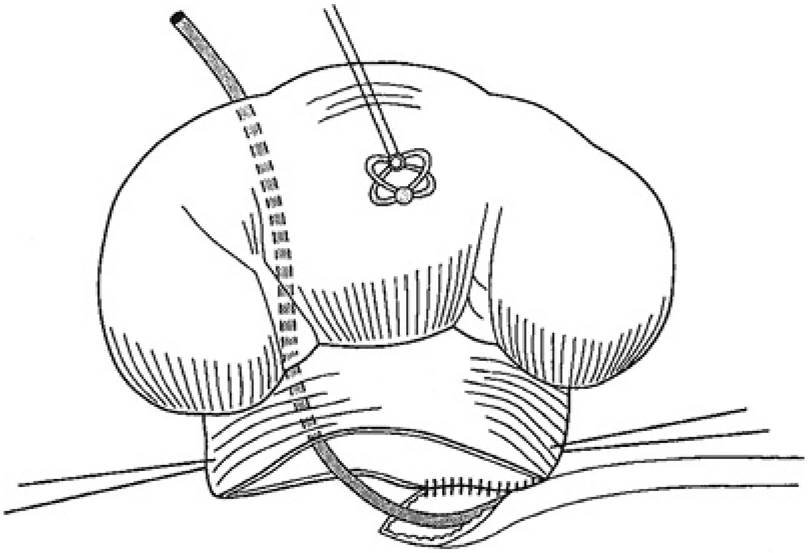
At this point, the authors consider a nephrostomy tube and ureteral stent (**Figure 67.6**). Neither is necessary for uncomplicated pyeloplasty, but intubation provides useful reassurance in difficult cases, floppy kidneys, revisions, or very redundant pelves. These can be ready-made or adapted from feeding tubes and small-caliber (10–12 Fr) Malecot catheters. In addition, a double-J ureteral stent may be used. The stent is advanced over a hydrophilic glidewire placed in an antegrade fashion. Position in the bladder can be confirmed by irrigating the Foley catheter with methylene blue-laced irrigation and observing reflux of the dyed irrigant from the ureter. The proximal curl is placed within the renal pelvis, and the anastomosis is completed.



67.6

Anastomosis

The anastomosis should begin at the caudal end, where the lower lip of the renal pelvis meets the spatulated apex of the ureter. The first suture (7/0, 6/0, or 5/0 in older children and adolescents) is placed at the apex and the next two sutures are positioned closely on either side. The length of the anastomosis should be ideally at least 1 cm and can be longer in older children. For infants and small children, the practical limits of the slack on the ureter and the reach of the pelvis often determine how long the anastomosis can be. A running or interrupted technique completes the process. Sutures should advance up the posterior wall (**Figure 67.7**) and then up the anterior wall, keeping placement of each even so that the tissue is equally distributed and bunching does not occur. When the end of the ureter has been reached, suturing continues until the pelvis is closed.



67.7

Fat wrap

A flap of fat freed from Gerota's fascia is carefully but loosely wrapped around the anastomosis and secured by stitching it to itself. A small open drain should be placed in the region of the repair but not overlying the anastomosis. Nephrostomy tubes and externalized stents are brought out through a separate stab incision. If none has been used, a drain is mandatory to manage potential urine leak.

Wound closure

The incision is closed in layers, with care taken to reapproximate the muscles. Local anesthetic may be infiltrated along the intercostal nerves for postoperative comfort. All tubes and drains should be securely sutured to the skin.

ROBOTIC-ASSISTED LAPAROSCOPIC PYELOPLASTY

Patient selection and contraindications

The robotic-assisted laparoscopic pyeloplasty has recently become a standard approach in pediatrics, particularly for older children. Performing the procedure through smaller incisions and with the assistance of a stereoscopic field of vision and wrist-joint articulation improves visualization and suturing capability for the surgeon while affording the patient a quick recovery and minimal postoperative pain requirements. A dismembered pyeloplasty can be successfully performed with good functional results even in the very young. However, this operation is recommended in smaller infants only if the surgeon is very adept at robotic surgery and is familiar with the anatomic and physiologic nuances of laparoscopy in small children.

Robot-assisted laparoscopic (RAL) pyeloplasty can also be performed on horseshoe kidneys, ectopic kidneys, and other kidneys with aberrant anatomy. Children presenting with intermittent obstruction with an intrarenal pelvis which only distends when obstructed present a special challenge because the renal pelvis will retract into the renal sinus once opened, unless the surgeon ensures that the pelvis is transfixed with a “hitch stitch.” Administration of a diuretic chased with a fluid bolus intraoperatively can facilitate distension of the renal pelvis for easier visualization and dissection.

Transperitoneal versus retroperitoneal approach

The retroperitoneal approach is popular for performing nephrectomies and heminephrectomies, having the supposed advantage of avoiding intraperitoneal adhesions. There is no statistical difference in the outcome of the intraperitoneal (transperitoneal) versus the retroperitoneal route. The authors prefer the transperitoneal route for pyeloplasty as it provides a much larger working space and better ergonomics, especially for fine intracorporeal suturing.

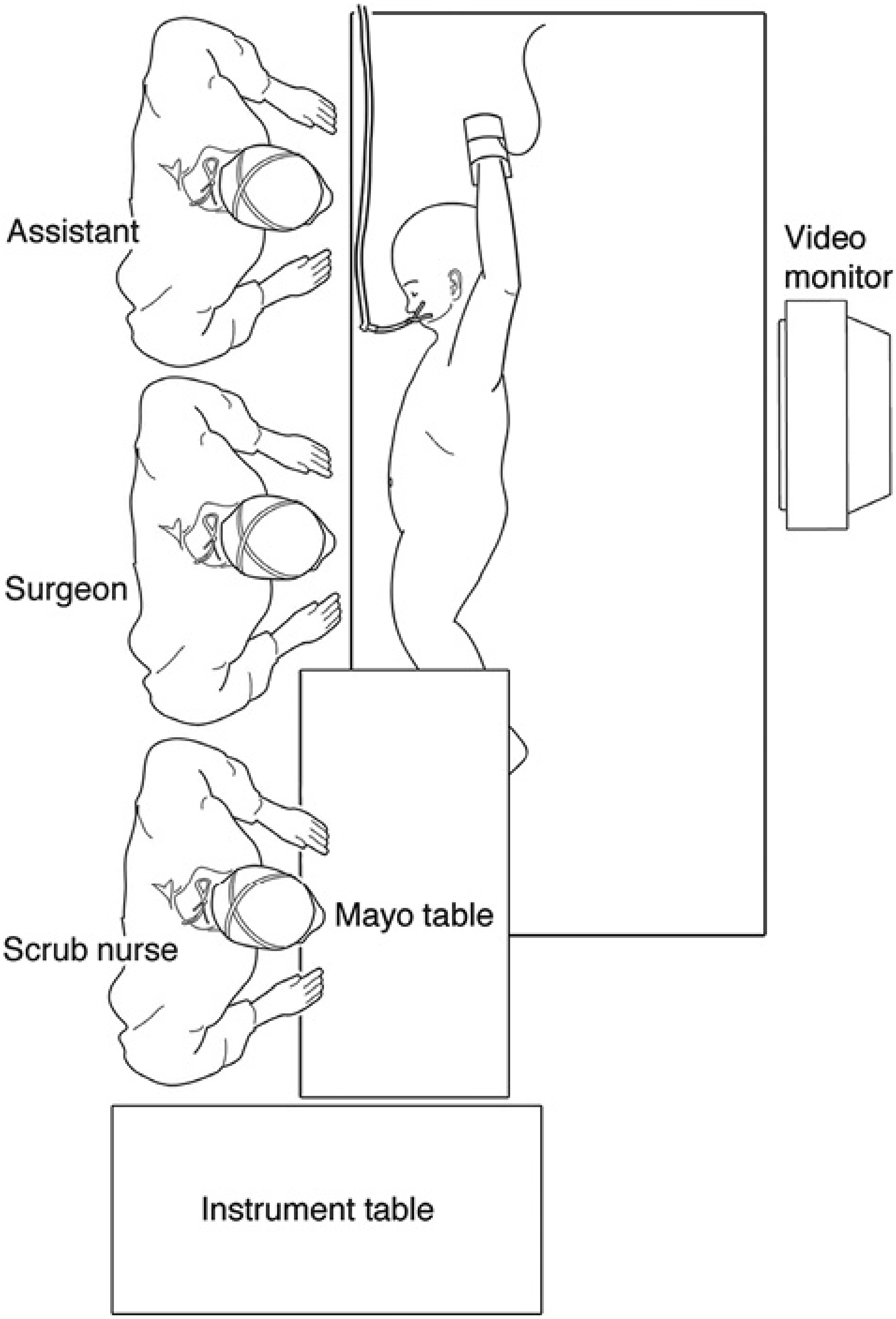
Instrumentation

It is not necessary to have an extensive range of robotic instruments. The authors prefer the standard 12 mm robotic camera, but an 8.5 mm robotic camera may be utilized. Preferences also include 5 mm robotic instruments, which allow for smaller incisions and limit need to close fascia in adolescents, as port-site hernias with 5 mm incisions are rare. Note that for younger patients fascia should be closed. A Maryland forceps, cautery hook, shears, and needle drivers are all that is required to complete the case successfully. The 5 mm instruments are limited by lack of bipolar cautery and monopolar cautery shears. For this reason, 8 mm instruments may be selected to broaden the range of instrumentation.

Preoperative preparation and general considerations

An enema to evacuate stool from the left colon could be considered, especially for left-sided UPJ obstruction, but this is not mandatory. While some authors advocate the insertion of a stent preoperatively, it has not always been found to be helpful as it only leads to complete decompression of the renal pelvis, making it more difficult to mobilize the pelvis and UPJ. The stent may also become entangled with the suture, making an already challenging task more difficult. It is the authors' preference to insert an antegrade stent as an intraoperative procedure, after completion of the posterior anastomosis. A transurethral Foley catheter should be inserted before the patient is positioned.

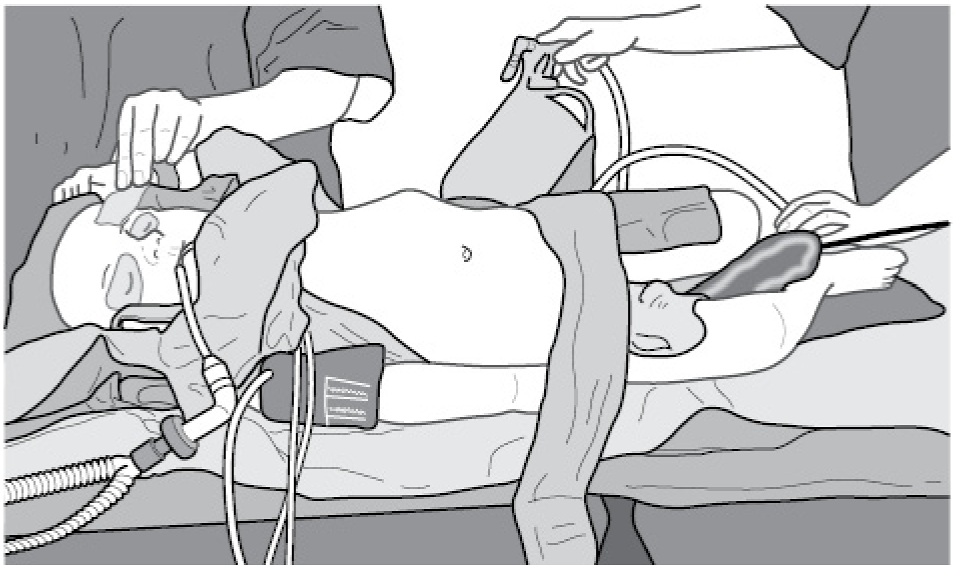
The patient, surgeon, assistant, and scrub nurse should initially be positioned as shown in the floor plan in **Figure 67.8**. This is the most ergonomic position.


Assistant
Video monitor
Surgeon
Mayo table
Scrub nurse
Instrument table

67.8

Patient positioning

A Foley catheter should be placed prior to positioning, and the anesthesia team should place an orogastric tube for bowel decompression. Complete paralysis is required throughout the case. The patient should be positioned at the edge of the table tilted at about 45-degree lateral decubitus. All dependent regions should be adequately padded, and an axillary roll should be placed beneath the dependent axilla. Flexing the table and raising the kidney rest helps open the abdomen to create more space for port placement. The patient should be secured to the table (**Figure 67.9**).

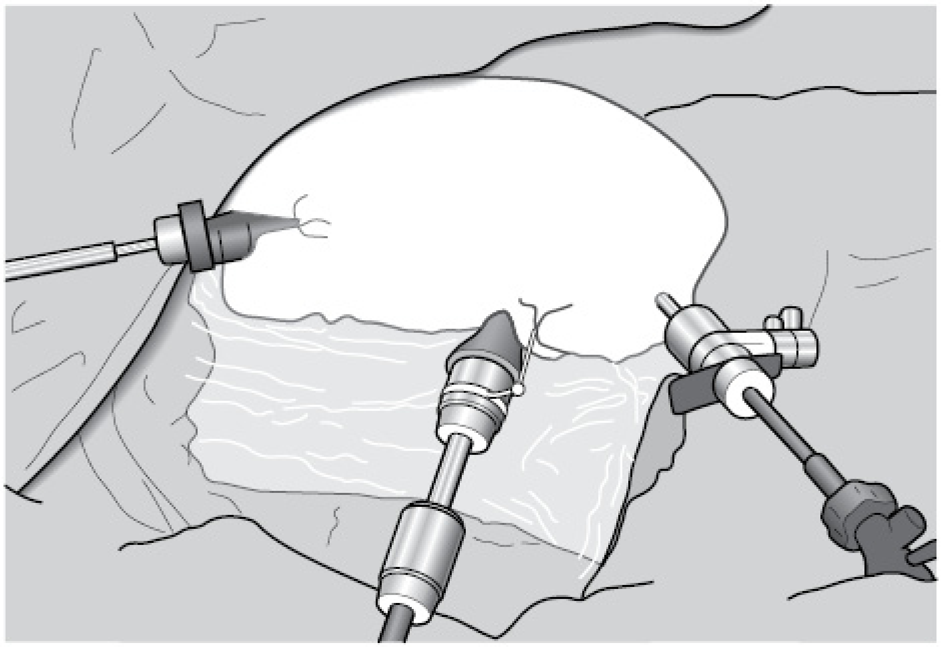


67.9

Incision and port placement

A supraumbilical curvilinear incision is then made down to the linea alba about 1 cm cephalad to the umbilical cicatrix, and the linea alba is grasped between two hemostats and incised. The peritoneum can be easily identified deep to this and opened. The robotic camera trocar is then advanced toward the lateral abdominal wall to avoid injury to the great vessels.

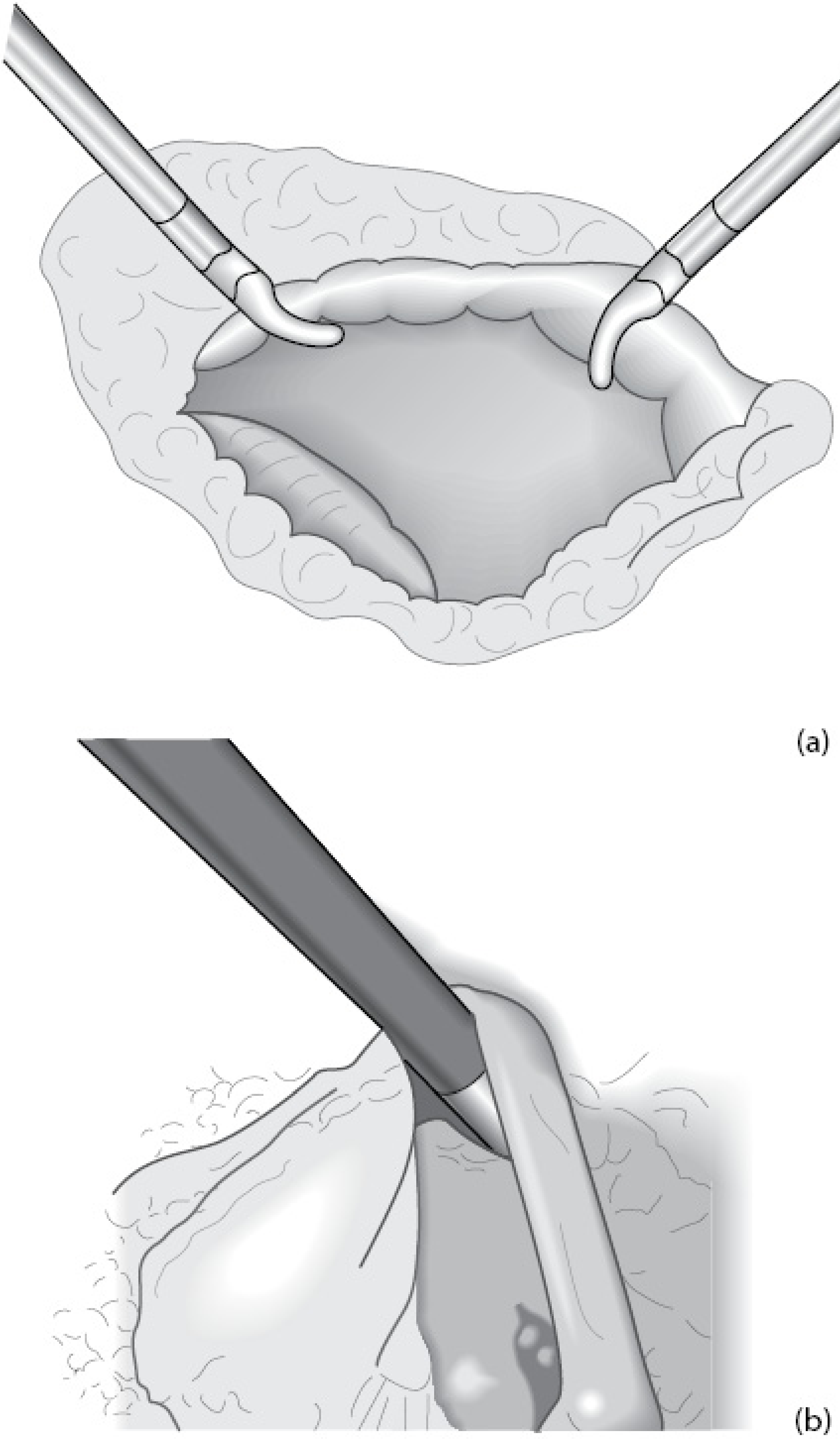
Port positions are critical for pyeloplasty. It is not possible to perform a satisfactory intracorporeal anastomosis if the needle driver is not in line with the intended line of anastomosis. Ideal robotic port placement is just lateral to the rectus abdominis, approximately 1–2 cm inferior to the costal margin and superior to the anterior superior iliac spine. A 5 mm or 8 mm robotic port may be selected. A 5 mm assistant port is best placed superior to the umbilicus. All ports should be separated by approximately 8 cm to avoid robotic arm collision (**Figure 67.10**).



67.10

Kidney mobilization and ureteral identification

The first step of the operation is to incise the peritoneum laterally and reflect the colon medially, away from the kidney and Gerota's fascia (**Figure 67.11a**). It is important to identify the correct tissue plane. The renal capsule is covered by a thin film of loose adventitia and it is important to be deep to this plane, keeping absolutely snug on the renal capsule. The plane is developed medially into the sinus where the renal pelvis should then be easily identified. The renal vein may be exposed during this maneuver.

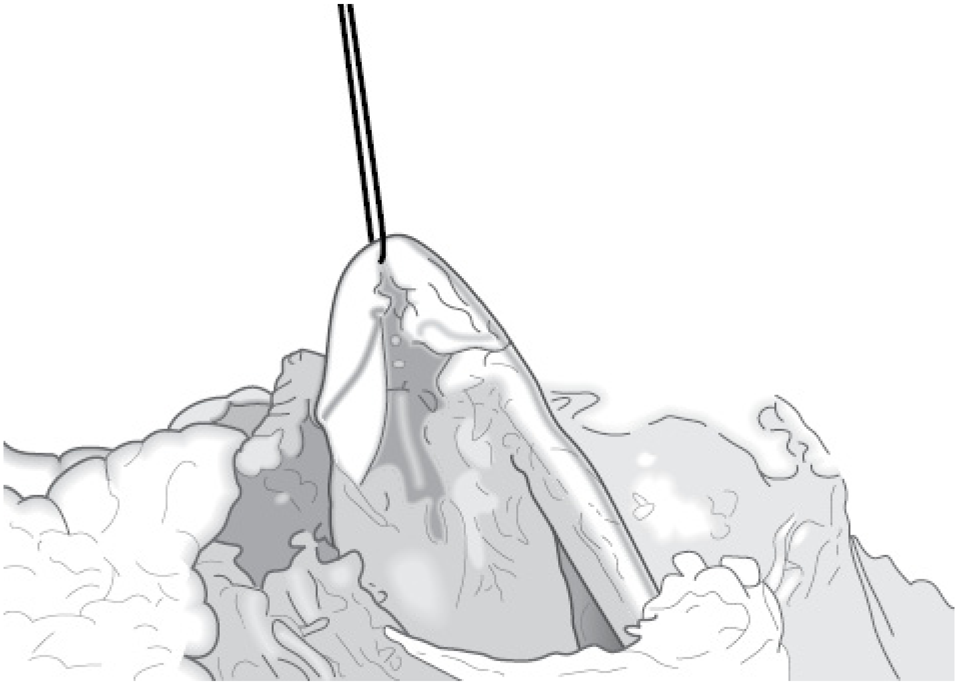


67.11a,b

The UPJ can usually be identified at this point. The gonadal vessels are usually located near the UPJ. Once the UPJ is identified, the ureter should be mobilized gently by lifting it out of its bed to develop a small window to hold it up. Minimal mobilization of the proximal ureter is performed to preserve the vascular supply. The renal pelvis is mobilized more extensively, sweeping the renal vessels toward the upper pole (**Figure 67.11b**). This is usually a bloodless procedure if you are in the correct plane. About 2 m of renal pelvis should be cleared above the upper limits of the intended pyelotomy. Care should be taken to preserve any crossing vessels and to dissect the UPJ until it can be flossed freely back and forth behind these vessels.

Renal pelvis isolation

At this point, a 0 monofilament suture on a either a large curved or straight needle is passed through the anterior abdominal wall at the mid- or posterior axillary line just below the costal margin and grasped from the inside with a robotic needle driver. The renal pelvis should be transfixed about 1–2 cm above the intended upper limits of the pyelotomy and the needle then passed through the abdominal wall near its insertion point. The renal pelvis can then be lifted out of its bed by this hitch stitch, displaying the UPJ in full view (**Figure 67.12**). A hemostat is applied to both ends of the hitch stitch on the outside to stabilize the renal pelvis for the duration of the pyeloplasty.



67.12

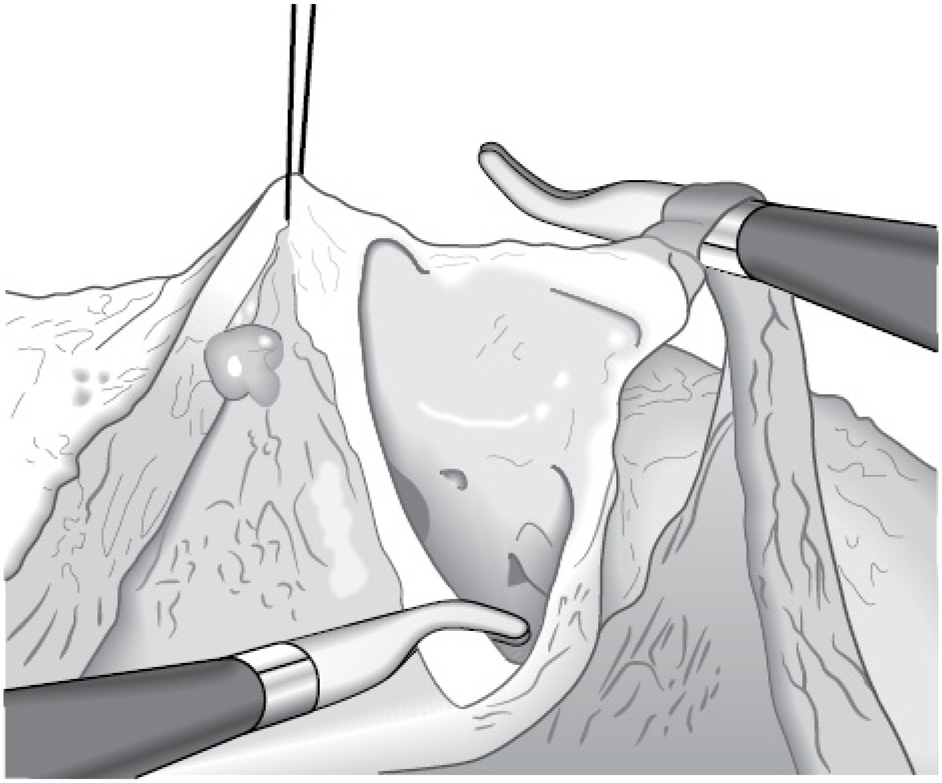
It is important to leave a good length of the suture on the outside, as it may be necessary to loosen the hitch stitch if the anastomosis is under tension. A second hitch stitch should not be placed on the proximal ureter as this makes for a more difficult anastomosis because of suture clash and may even distract the ureter from the renal pelvis.

It is also important at this stage to spend a few minutes to clear the renal pelvis and proximal ureter of any loose connective tissue before dividing the UPJ, otherwise the ureter will retract within the sleeve of connective tissue, which will make for a difficult anastomosis. Follow the proximal ureter down to ensure that the obstruction is not caused by a crossing lower-pole vessel.

UPJ division

With the UPJ now stabilized by the hitch stitch, the most dependent part of the pelvis is identified and opened. Only the anterior wall of the renal pelvis is then divided along the entire length of the intended pyelotomy

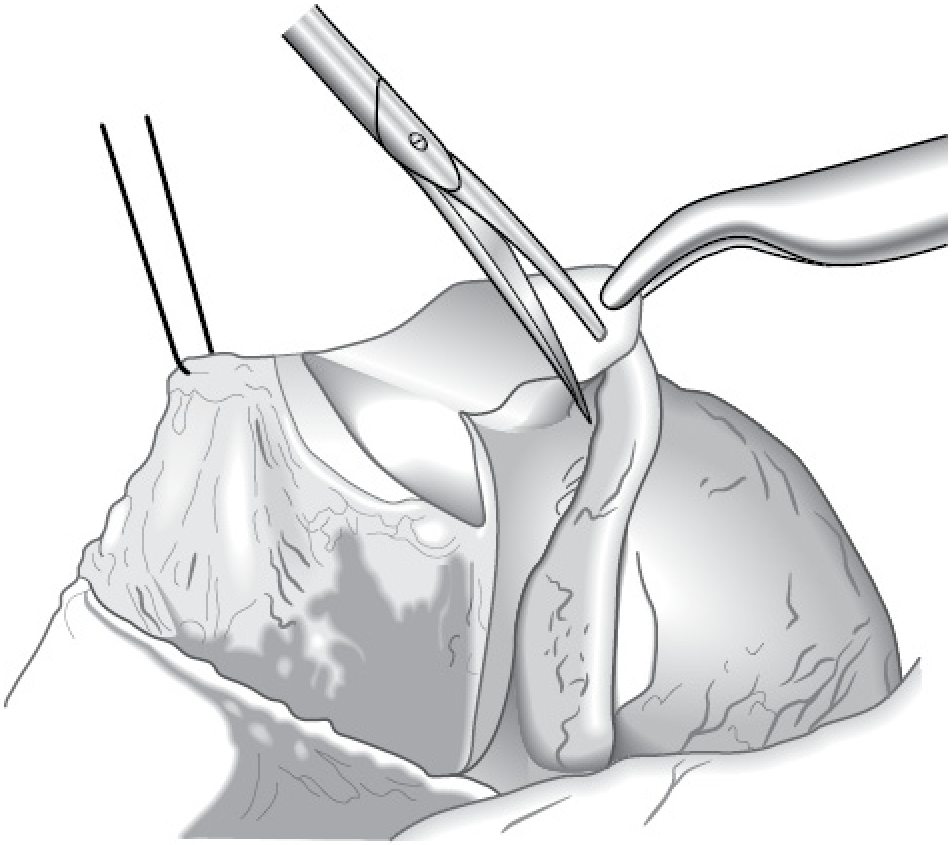
(**Figure 67.13**). The posterior wall of the pyelotomy, especially the superior portion, should be left intact at this point.



67.13

Ureteral spatulation and UPJ excision

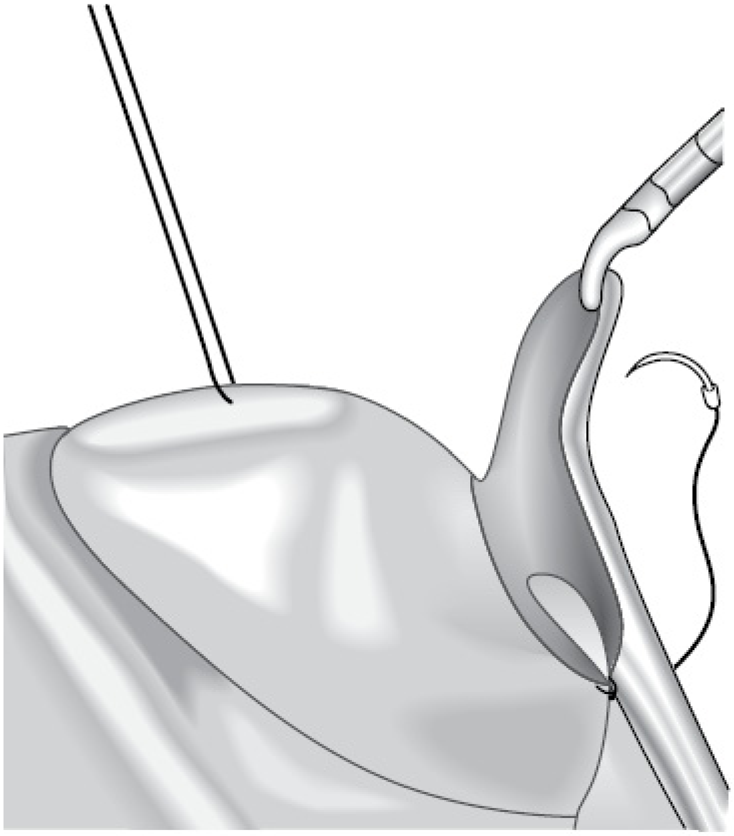
The UPJ should not be completely dismembered, as the attached posterior wall makes it much easier to spatulate the ureter since the ureter can be stabilized by applying countertraction on the partially divided pelvis (**Figure 67.14**). Once the spatulation is complete, the ureter may then be dismembered completely.



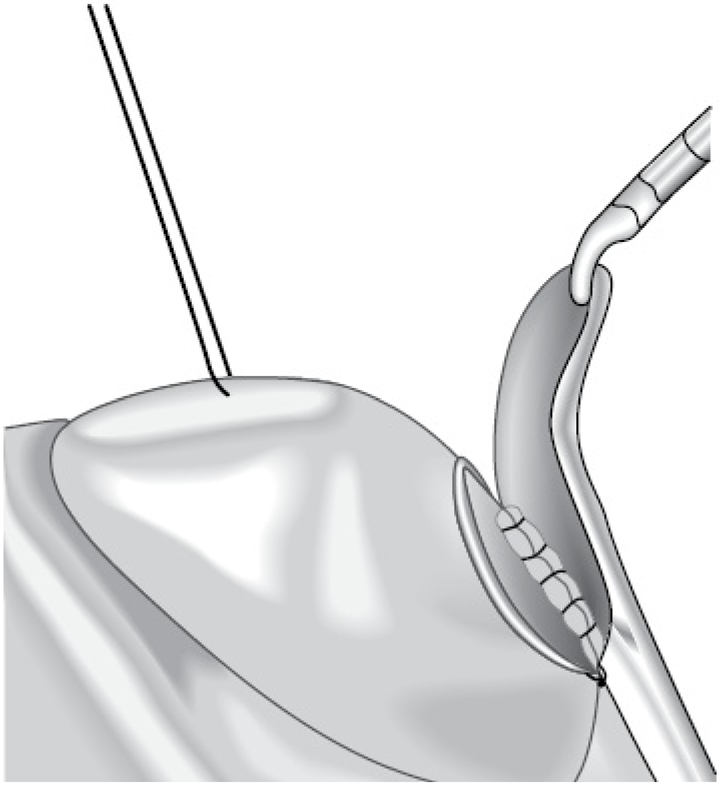
67.14

Anastomosis

The anastomosis is begun with a 5/0 or 6/0 continuous absorbable monofilament suture. The first suture should be placed at the angle of the spatulated ureter, which is then approximated to the most dependent part of the renal pelvis as in open pyeloplasty (**Figure 67.15a**). The redundant pelvic tissue can be grasped with impunity to bring the tissue to the ureter during the anastomosis. Only the ureteral adventitia should be manipulated when handling the ureter to avoid harm to the mucosa. The posterior anastomosis is completed as a continuous suture. The anastomosis should then be inspected internally by opening up the ureter (**Figure 67.15b**).

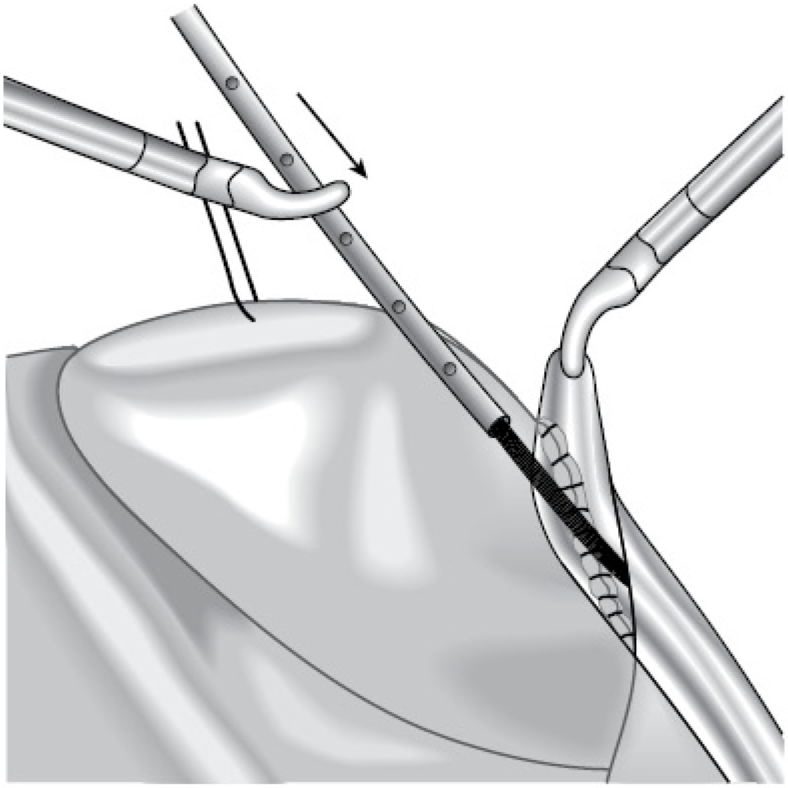


(a)

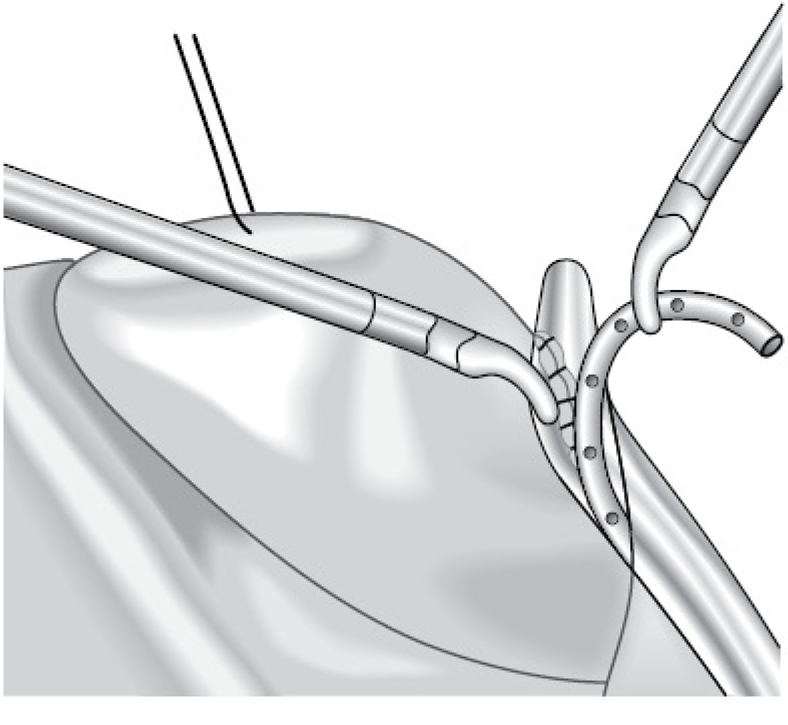


67.15a,b

The antegrade stent should then be inserted at this point. A 14 gauge angiocath is inserted through the anterior abdominal wall just in front of the hitch stitch and in line with the ureter. A hydrophilic glidewire preloaded with a double-J ureteral stent is then advanced via the angiocath down the ureter (**Figure 67.16a**). Once the marking at the proximal curl is visualized at the UPJ, the wire is removed, and the proximal curl is situated within the renal pelvis (**Figure 67.16b**). Position can be confirmed by irrigating the Foley with methylene blue-laced saline and visualizing reflux at the anastomosis (**Figure 67.16c**) or by obtaining an abdominal X-ray postoperatively.



(a)



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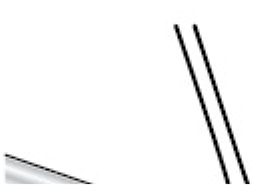
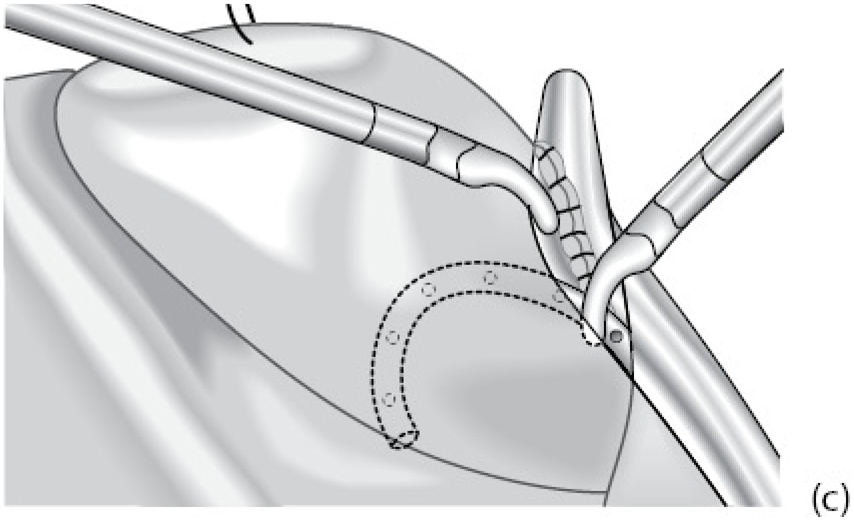
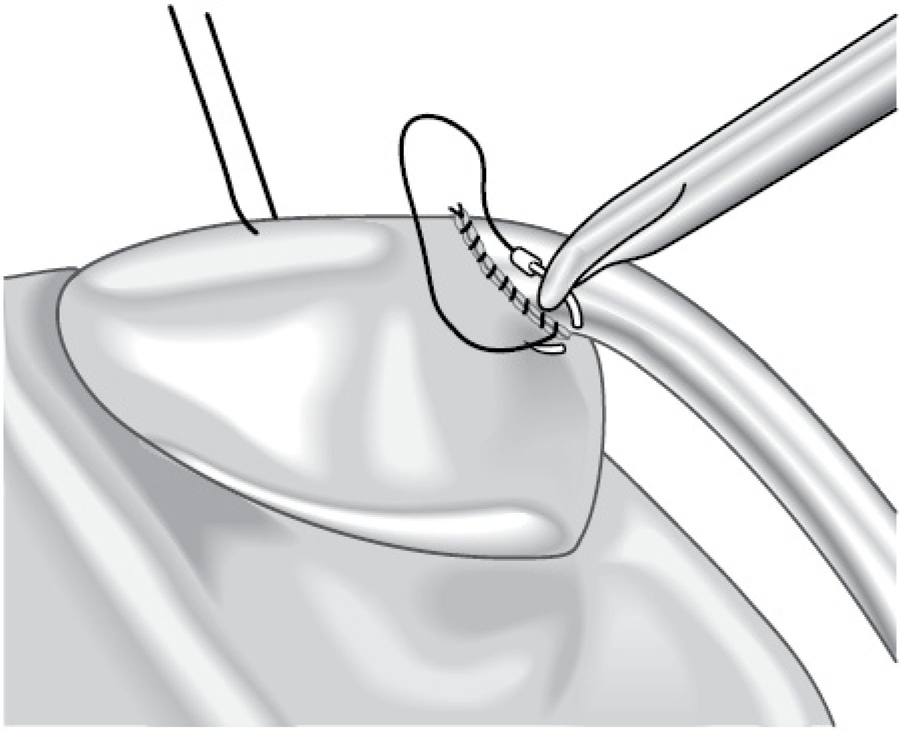




67.16a–c

The anterior anastomosis is completed with a 5/0 or 6/0 continuous absorbable monofilament suture, beginning again at the inferior aspect of the ureteral spatulation and pyelotomy (**Figure 67.17**). The anastomosis is then inspected after the hitch stitch is removed and the pelvis and ureter returned to its bed, ensuring that it is not kinked.



67.17

Wound closure

All port sites are closed at the fascial level with 3/0 absorbable monofilament suture. In adolescents, risk of incisional hernias is less for port sites less than 7 mm, so skin closure only could be considered. Skin is closed at each port site in a subcuticular fashion with 5/0 absorbable monofilament suture, and each wound is dressed with topical skin adhesive.

Points of technique

Patients presenting with intermittent UPJ obstruction with a predominantly intrarenal pelvis present a special challenge as the intrarenal pelvis can severely restrict the surgeon's ability to manipulate the laparoscopic instruments. It requires the patient to have a fluid load and furosemide to produce maximum diuresis. The surgeon should wait for maximal distention of the renal pelvis before placing the hitch stitch to lift the renal pelvis away from the hilum. Under no circumstances should the UPJ be divided before the renal pelvis is transfixed, as this will result in the pelvis retracting into the depths of the hilum, making suturing impossible.

Crossing lower-polar vessels are easily identified laparoscopically without the need for CT or MRI. If the pathology is an aberrant lower-pole vessel, it will be necessary to completely dismember the ureter from the pelvis in order to transpose the vessels.

On completion of pyeloplasty, aspirate any urine that has drained into the surgical field and inspect for hemostasis. The colon is then remanipulated into its original position. The peritoneal incision will then be completely covered by the colon, leaving no raw surfaces. There is no need for a perinephric drain. Left-sided pyeloplasties may be completed through a transmesenteric approach provided there is a large enough window in the mesentery to expose the UPJ without compromising any mesenteric vessels. The mesenteric window does not require closure upon completion of the case.

POSTOPERATIVE CARE

The patient may be advanced quickly to an age-appropriate regular diet immediately following surgery, particularly in the setting of a retroperiteoneal approach. The urethral catheter is removed the following morning. For open pyeloplasty, if the flank remains dry, the drain may be removed in the next day or so. If the repair is intubated with an external stent and nephrostomy tube, the tube should be left in place for 1–3 weeks, after which the stent may be removed and a nephrostogram is performed. If no drainage is seen flowing down the ureter, the tube should be left to drainage for a further 2 weeks and the test repeated. If contrast flows across the repair, the tube may be removed. Indwelling double-J ureteral stents may be left in place for 4–6 weeks and require general anesthesia for cystoscopic removal. Follow-up renal US is arranged 3–6 months after surgery. The decision to obtain a postoperative renal scan depends on the patient's clinical course and degree of hydronephrosis following surgery.

COMPLICATIONS

Intraoperative complications include bleeding possibly requiring transfusion, damage to intraperitoneal structures, entry into the thoracic cavity requiring chest tube placement, the need to convert to an open approach in the setting of a robotic-assisted laparoscopic case, or the need to perform a nephrectomy. During the postoperative course, the patient could develop a UTI or wound infection. Urine leak or abscess warranting drainage may arise. The principal long-term complication is stenosis of the anastomosis. Fortunately, this is rare and 90–95% of cases are successful. Although the repaired kidney may never look normal on US due to persistent hydronephrosis, it should function and drain normally on follow-up diuretic renal scan if performed.

LAPAROSCOPIC TRANSPOSITION OF LOWER-POLE CROSSING VESSELS: THE "VASCULAR HITCH"

Crossing lower-pole renal vessels have been implicated in the etiology of UPJ obstruction in up to 30% of cases. Conventional surgical treatment in these cases is standard dismembered pyeloplasty, Hellström's nephroplication, or transposition of lower-pole crossing vessels, which may be performed laparoscopically in selected patients. This may be an attractive surgical alternative, as it eliminates the need to violate the collecting system, thus avoiding the potential anastomotic complications, and is also less challenging than performing fine intracorporeal suturing.

The crossing lower-pole vessels should be plicated to the renal pelvis cephalad from the UPJ by imbricating the redundant pelvis over the vessels with two or three absorbable sutures.

SPECIAL SITUATIONS

Combined UPJ obstruction and vesicoureteric reflux

Patients who present with both vesicoureteric reflux and ipsilateral UPJ obstruction can be complicated. High-grade vesicoureteric reflux can distort the appearance of the UPJ. One should ask if the dilation of the renal pelvis is due to the reflux, or whether it is a separate concern. In these cases, the surgeon should first determine if there is truly an obstruction. Diuretic renal scintigraphy will rule out most cases of pseudo-UPJ obstruction. When obstruction has been established, the UPJ obstruction should be repaired first, and the reflux addressed later. Repairing the reflux first risks making the obstruction worse and jeopardizes the kidney.

Duplex systems

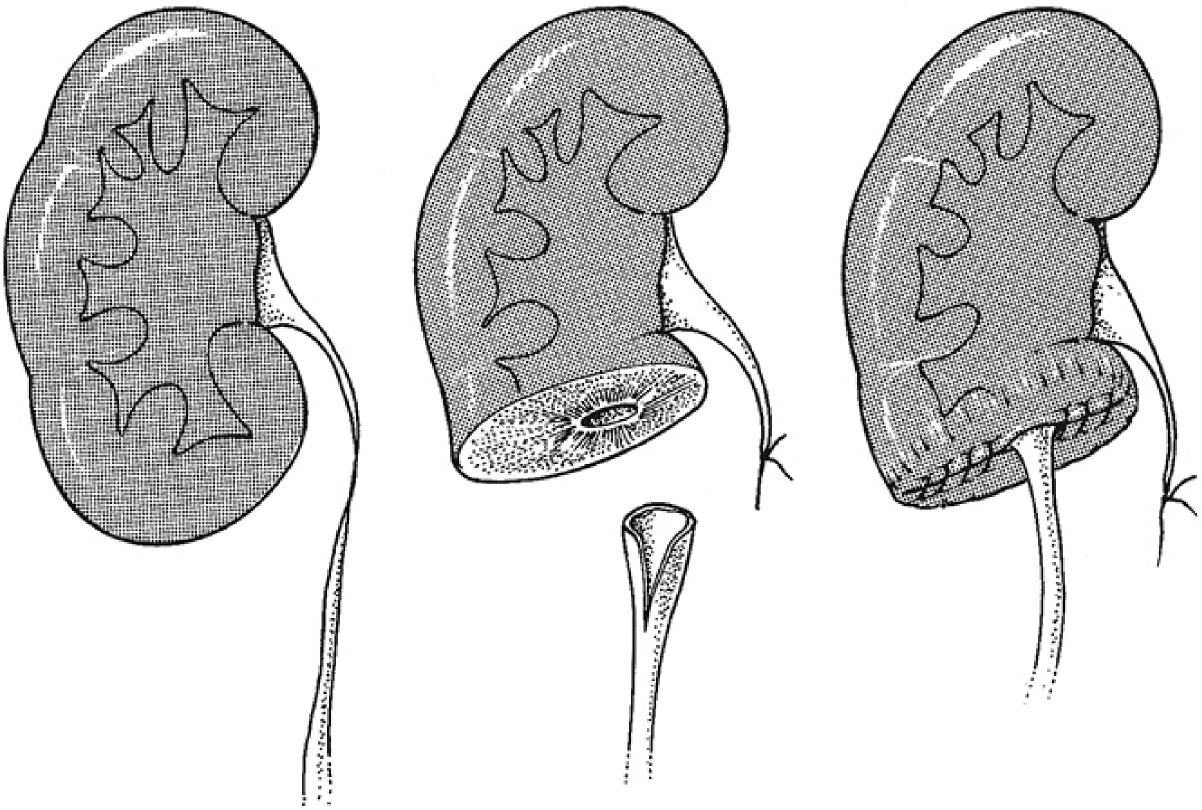
Obstruction is most commonly found in the lower-pole moiety of duplicated systems. The open pyeloplasty technique described above may be used, but the surgeon must be careful to identify and isolate the appropriate ureter. If a particularly long obstruction is found, ureteropyelostomy or a pyelopyelostomy may be advantageous.

Horseshoe kidneys

In horseshoe kidneys, the ureter often inserts high into the renal pelvis; a longer, more caudally placed anastomosis is needed. Division of the isthmus is usually not necessary. A Y–V flap advancement style of pyeloplasty may be useful in some anatomic configurations, but a dismembered pyeloplasty can usually be done.

Ureterocalycostomy

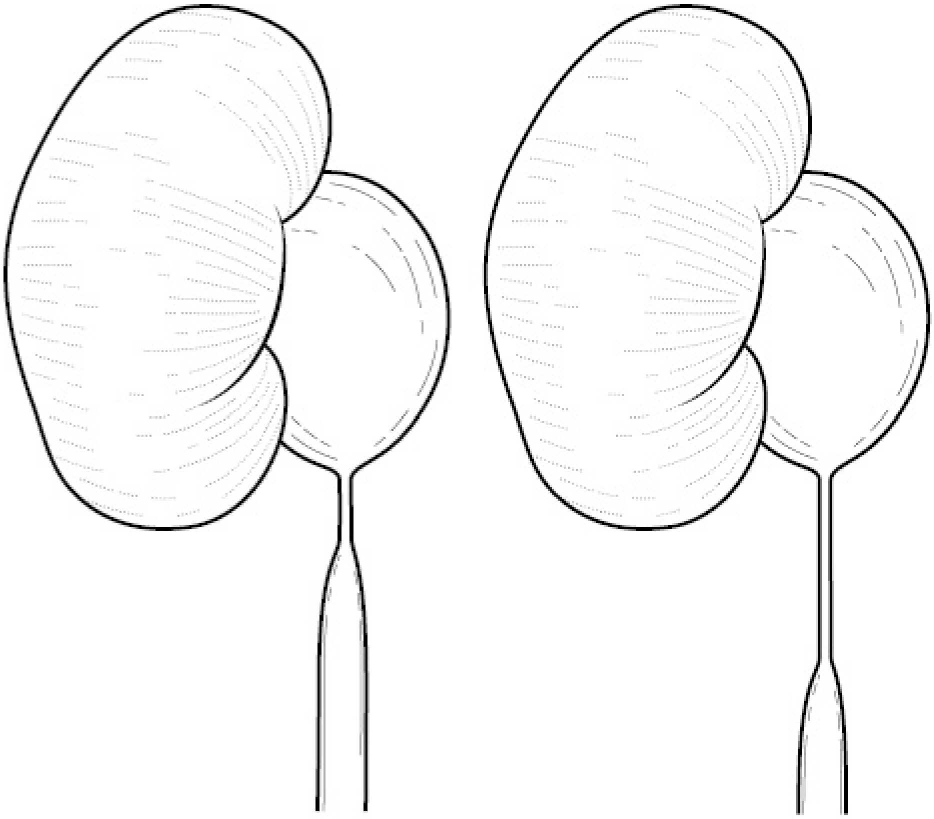
If the renal pelvis is too small, intrarenal, or too fibrotic to achieve dependent drainage, a ureterocalycostomy may be necessary. The principal features of this rare procedure are the selection of the most dependent calyx and a thorough amputation of the lower-pole of the kidney to prevent postoperative compression (**Figure 67.18**).



67.18

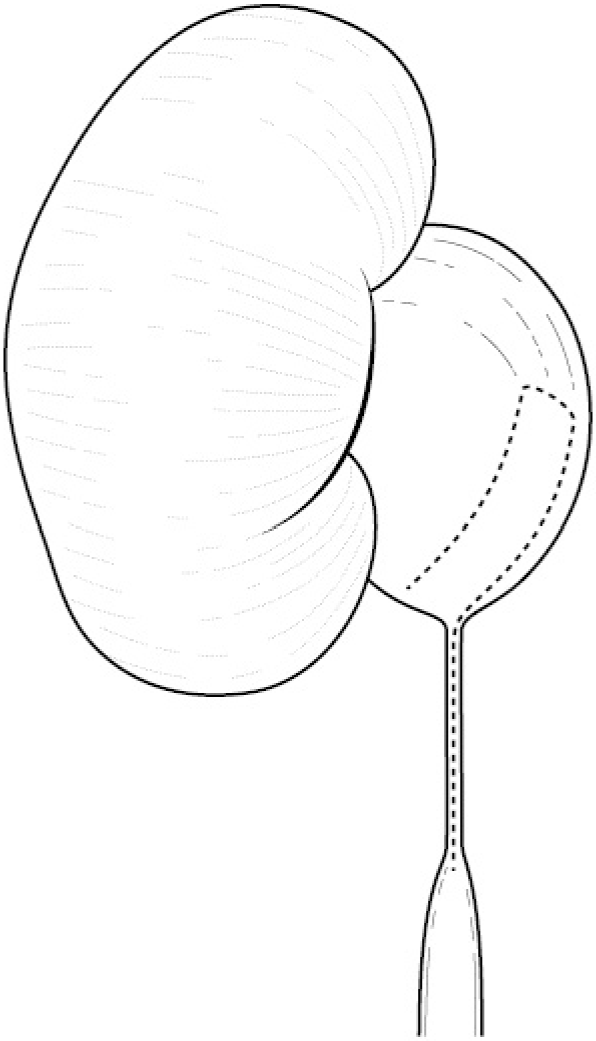
Flap pyeloplasty

Usually, the area of stenosis is short and the pelvis and normal ureter are in very close approximation (**Figure 67.19**). If there is a long stenotic ureteral segment, before disconnecting the ureter the surgeon should be sure that an anastomosis can be done without tension. If the stenotic segment is 2 cm or longer, attempts should be made to fully mobilize the kidney and ureter. If an anastomosis cannot be done without tension, a flap pyeloplasty should be considered.



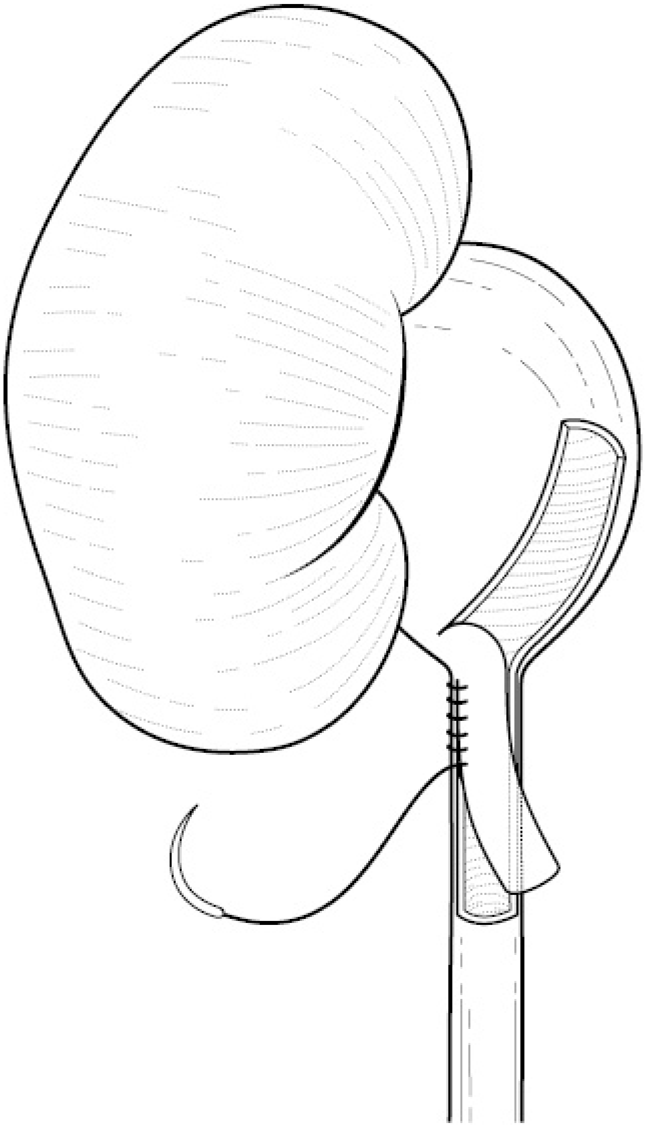
67.19

The stenotic segment and the flap should be carefully measured and marked out so that they will closely match. The incision should completely open up the stenotic portion. The flap should be equal to the length of the segment. The flap can be straight or can curve around the contour of the pelvis as needed (**Figure 67.20**). Care should be taken to ensure the base of the flap is wide enough to provide adequate blood supply to the tip. The width of the flap should be about the circumference of the normal ureter. Stay sutures should be used to help orient and control the flap and ureter. They will also help obviate the need to pick up the ureter and flap with forceps.



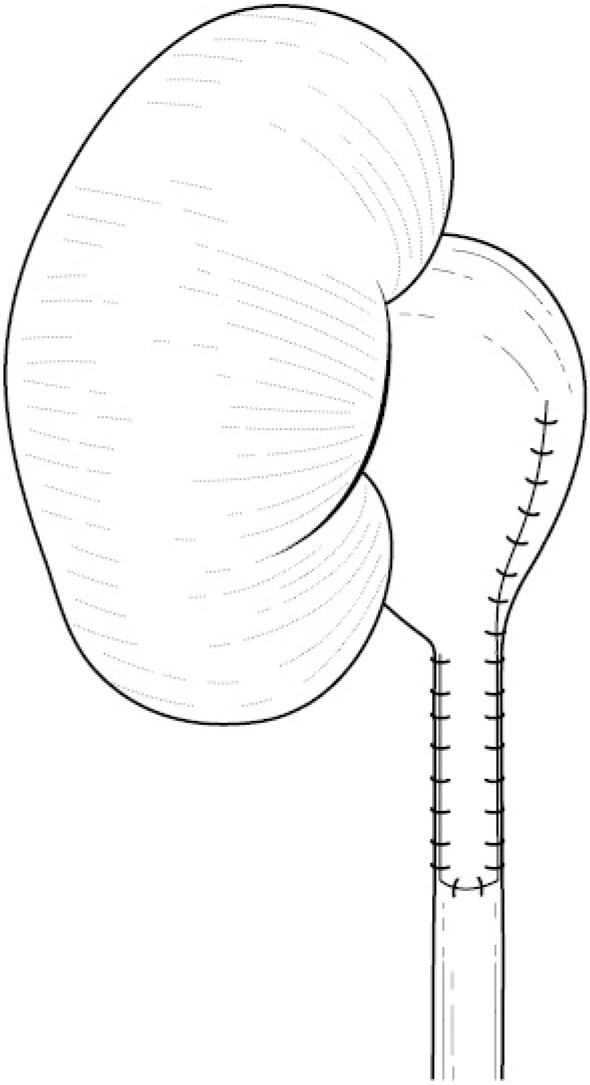
67.20

The flap is incised using a fresh hooked blade (a no. 12 scalpel) or sharp dissection scissors. The flap is rotated down and the lateral edge carefully sewn to the incised ureter (**Figure 67.21**). Interrupted or running sutures can be used. If a running anastomosis is used, consideration should be given to placing an interrupted suture at the distal end. This will help line up the edges to prevent bunching and spiraling of the running stitch.



67.21

The anterior edge of the flap should be closed with a similar stitch (**Figure 67.22**). Prior to this, placement of a stent and nephrostomy tube are advisable in addition to the usual Penrose drain. The stent should be left in for 1–3 weeks.



67.22

Ileal ureter

In some situations, the gap is too long to bridge by mobilizing the kidney and distal ureter. This is usually observed in patients who have already undergone extensive surgery or trauma resulting in a long atretic ureter. In such patients, an interposition of a segment of prepared ileum is an option. On the right side, an appendiceal interposition may prove to be very useful. This interposition should be placed so that the normal peristaltic course is directed toward the bladder (isoperistaltic).

FURTHER READING

Anderson JC, Hynes W. Retrocaval ureter: A case diagnosed pre-operatively and treated successfully by a plastic operation. *Br J Urol* 1949; 21: 209–14.

Boysen WR, Gundeti MS. Robot-assisted laparoscopic pyeloplasty in the pediatric population: A review of technique, outcomes, complications, and special considerations in infants. *Pediatr Surg Int* 2017; 33(9): 925–35.

Culp OS, DeWeerd JH. A pelvic flap operation for certain types of ureteropelvic obstruction: Observations after two years' experience. *J Urol* 1954; 71: 523–9.

Gundeti MS, Reynolds WS, Duffy PG, Mushtaq I. Further experience with the vascular hitch (laparoscopic transposition of lower pole crossing vessels): An alternate treatment for pediatric ureterovascular ureteropelvic junction obstruction. *J Urol* 2008; 180: 1804–32.

Hendren WH, Radhakrishnan J, Middleton AW Jr. Pediatric pyeloplasty. *J Pediatr Surg* 1980; 15: 133–44.

Kavoussi LR, Meretyk S, Direks SM, et al. Endopyelotomy for secondary ureteropelvic junction obstruction in children. *J Urol* 1991; 145: 345–9.

Rassweiler JJ, Teber D, Frede T. Complications of laparoscopic pyeloplasty. *World J Urol* 2008; 26: 539–47.

Scardino PL, Prince CL. Vertical flap ureteropelvioplasty. *South Med J* 1953; 46: 325–31.

Silay MS, Spinoit A, Bogaert G, et al. Imaging for vesicoureteral reflux and ureteropelvic junction obstruction. *Eur Urol Focus* 2016; 2(2): 130–8.

Tubre RW, Gatti JM. Surgical approaches to pediatric ureteropelvic junction obstruction. *Curr Urol Rep* 2015; 16: 72.

68

Nephrectomy and partial nephrectomy

IMRAN MUSHTAQ, ALBERTO MANTOVANI, AND JUDY HUNG WING SUET

OPEN NEPHRECTOMY

HISTORY

The first successful nephrectomy was performed in 1869 by the German surgeon Gustav Simon for a 46-year-old washerwoman who had undergone a hystero-ophorectomy and developed refractory ureteral obstruction due to adhesions. Simon, who had proved the compatibility with life of single-kidney animal models, decided to remove the kidney. The operation was performed through a flank incision with retroperitoneal approach and completed in 40 minutes with only 50 mL blood loss.

The first partial nephrectomy (heminephrectomy) is widely attributed to Czerny in 1889, and in 1884 Wells performed a partial nephrectomy to remove a perirenal lipoma. In the 1940s partial nephrectomy became a practical method for treating localized tuberculous disease of the kidney.

PRINCIPLES AND JUSTIFICATION

Indications

The most common indications for nephrectomy or nephroureterectomy in modern pediatric practice include:

- Congenital poorly functioning dysplastic kidney
- Reflux-associated nephropathy with loss of function (LOF)

Multicystic dysplastic kidney (MCDK), previously considered an indication for nephrectomy, is now widely managed conservatively unless the MCDK is particularly large and fails to involute or is associated with hypertension.

Less common indications include the following:

- Ureteropelvic junction (UPJ) obstruction with LOF
- Xanthogranulomatous pyelonephritis causing significant LOF
- Congenital nephrotic syndrome causing intractable protein loss
- Pretransplant nephrectomy of native kidney(s) in selected patients

Residual kidney function of around 10% is generally considered the cut-off for considering nephrectomy. However, this is a source of debate as many conditions might benefit from the preservation of any residual function (e.g. patients with metabolic predisposition to nephrolithiasis or patients with significant chronic kidney disease).

The most common indication for partial nephrectomy in a child is a poorly functioning upper moiety of a duplex kidney. In such instances, the upper pole is usually dilated due to an obstructing ureterocele or there may be a small dysplastic upper moiety with ectopic insertion of the distal ureter, usually in girls. Less frequently, a lower-pole partial nephrectomy is required when there is reduced lower moiety function with significant vesicoureteric reflux (VUR) and/or recurrent urinary tract infections (UTIs).

PREOPERATIVE ASSESSMENT

Preoperative preparation always includes a detailed review of all radiological investigations. This includes the most recent renal ultrasound, a DMSA and/or MAG3 scan and in most cases, a micturating cystourethrogram. The information gained from these investigations confirms the correct side for surgery and allows for planning the length of the incision, the extent of ureterectomy, and whether the ureter requires ligation or transection alone.

OPERATIONS

Approach

There are several approaches for renal surgery in children. The flank or lateral approach is derived from experience in adults, but most pediatric surgeons and urologists favor the anterior muscle-splitting extraperitoneal approach. This approach gives excellent exposure of the kidney through a relatively small skin incision and is suitable for nephrectomy and partial nephrectomy for benign diseases. In addition, it gives a very acceptable cosmetic outcome.

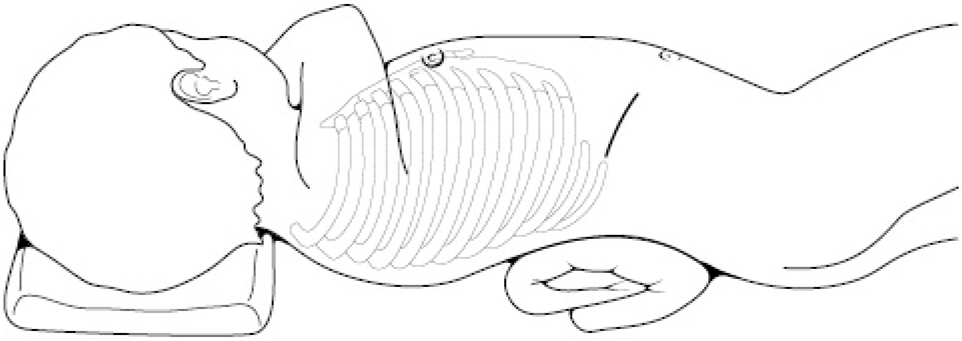
The transperitoneal (TP) approach, favored for oncological procedures (see Chapter 62), should be considered in cases of xanthogranulomatous pyelonephritis or when it is anticipated there will be dense scarring around the kidney and renal hilum. This approach has the advantage of providing rapid access to the renal hilum and major vessels. Inevitably, the choice of approach will also be influenced by the surgeon's experience and training.

In this chapter, only the anterior muscle-splitting extraperitoneal approach will be discussed.

Nephrectomy

POSITION OF PATIENT

The patient is positioned supine with the affected side raised with a cotton wool or gel-filled support. The ipsilateral arm is placed across the chest and the head is turned to the contralateral side (**Figure 68.1**). The incision for the anterior muscle-splitting approach extends medially from the tip of the twelfth rib toward the umbilicus. The length of the incision ranges from 3 cm in infants to 5–6 cm in older children. The planned incision should be marked with an indelible surgical marker prior to skin preparation.



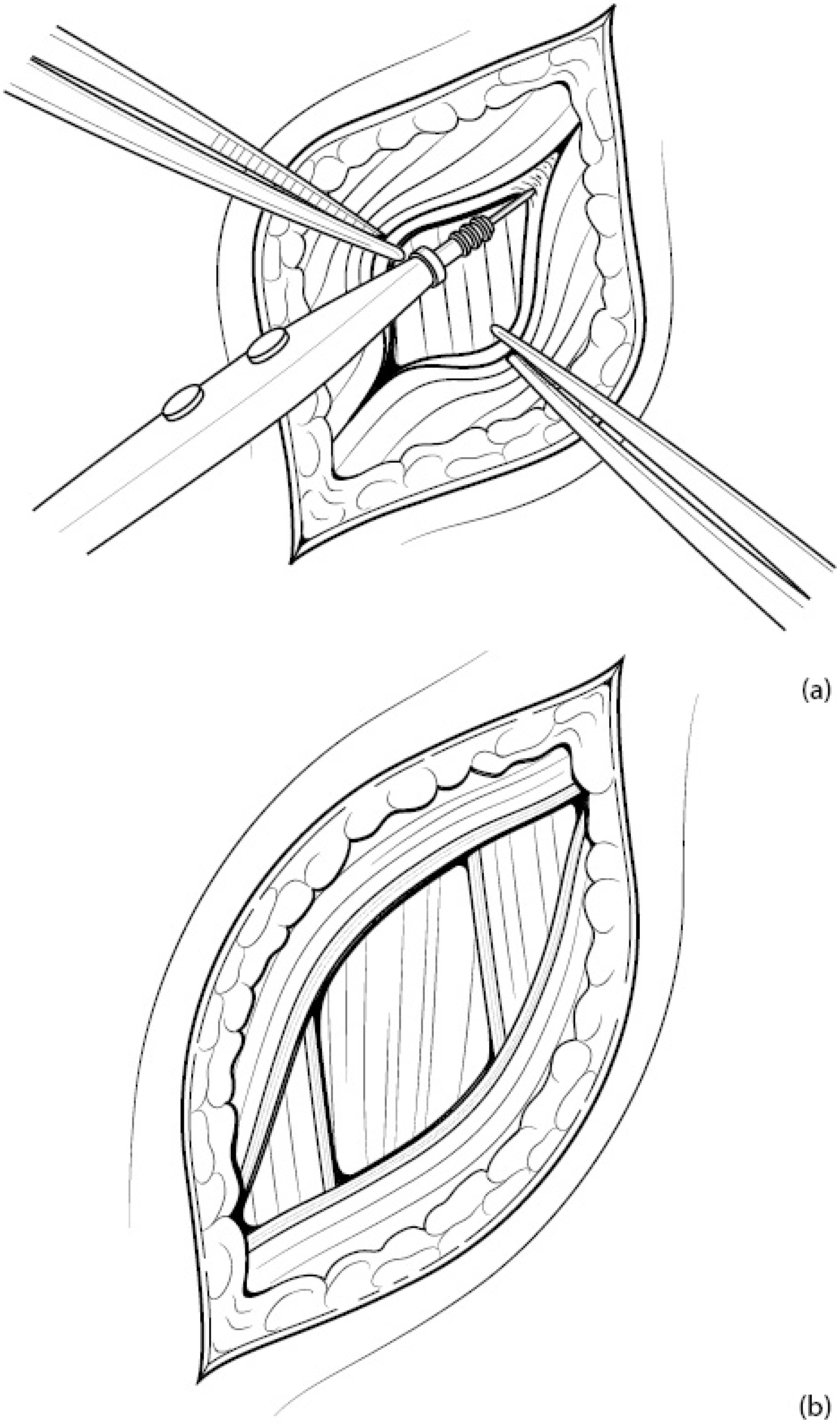
68.1

INCISION

The skin incision is made along the line previously marked. Monopolar diathermy is used to deepen the incision, dividing Scarpa's fascia and exposing the underlying external oblique muscle. Using Kilner cat's paw retractors to lift the edges of the skin and subcutaneous tissue, the wound is undermined in a circumferential manner to expose a broad area of the external oblique muscle, particularly in a posterior direction. This maneuver is essential for achieving good exposure of the kidney through a relatively small skin incision.

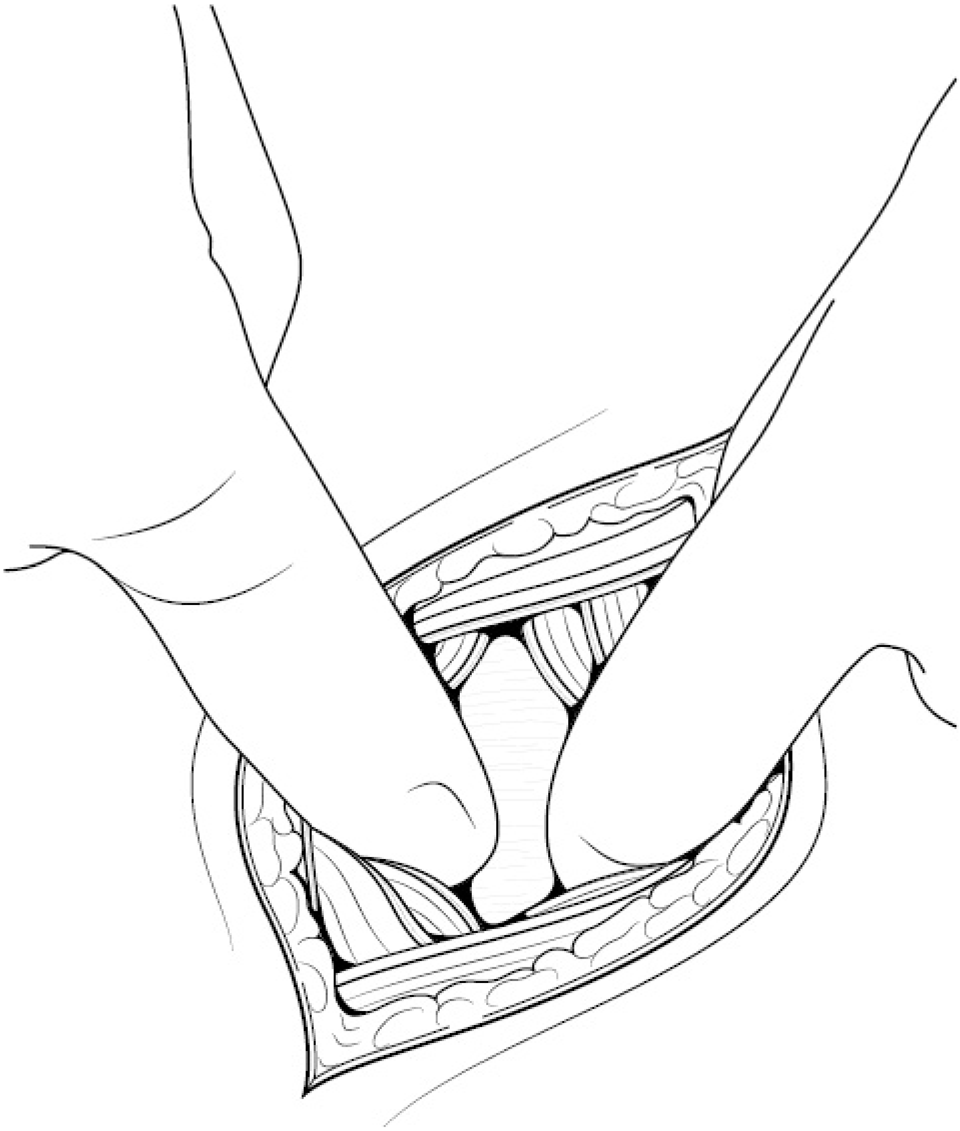
PROCEDURE

Monopolar diathermy is used to split the external and internal oblique muscle layers, along the line of the muscle fibers, to expose the underlying transversus abdominis muscle (**Figure 68.2a**). Once again, the muscle layers are undermined to expose a broad area of muscle (**Figure 68.2b**). Care is taken to identify and preserve the neurovascular bundles. The space between external and internal oblique muscles is usually easier to create compared to the space between internal oblique and transversus abdominis muscle.



68.2a,b

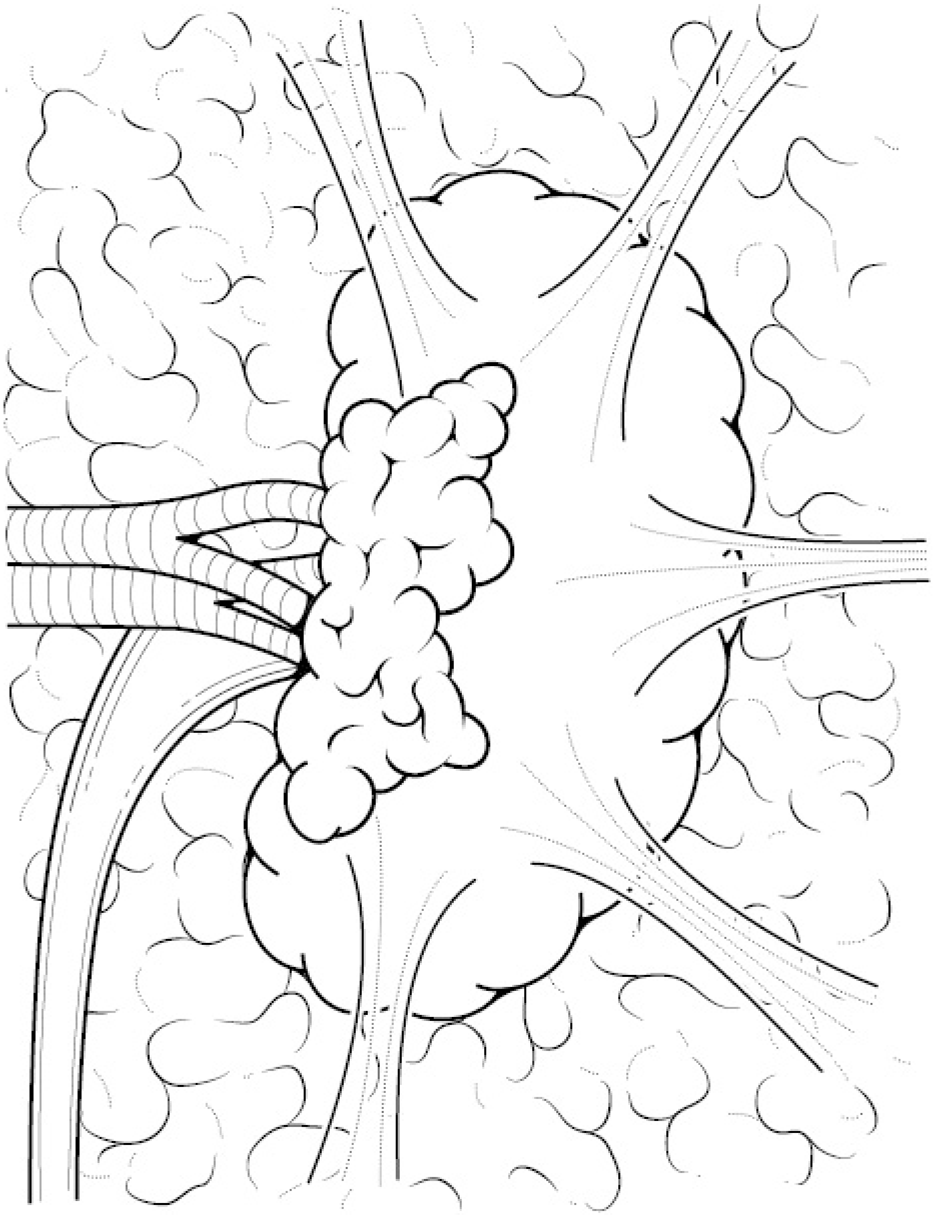
Commencing posteriorly to avoid entering the peritoneal cavity, the transversus abdominis muscle is split gently with scissors and retractors. Using the index fingers of both hands, the peritoneum is swept medially, moving in a posterior direction until the vertebral column can be palpated (**Figure 68.3**). A self-retracting or hand-held retractor can be used to display the kidney beneath Gerota's fascia. Gerota's fascia is entered with scissors and the opening enlarged with the index fingers of both hands. The surface of the kidney should be easily identifiable beneath the perinephric fat.



68.3

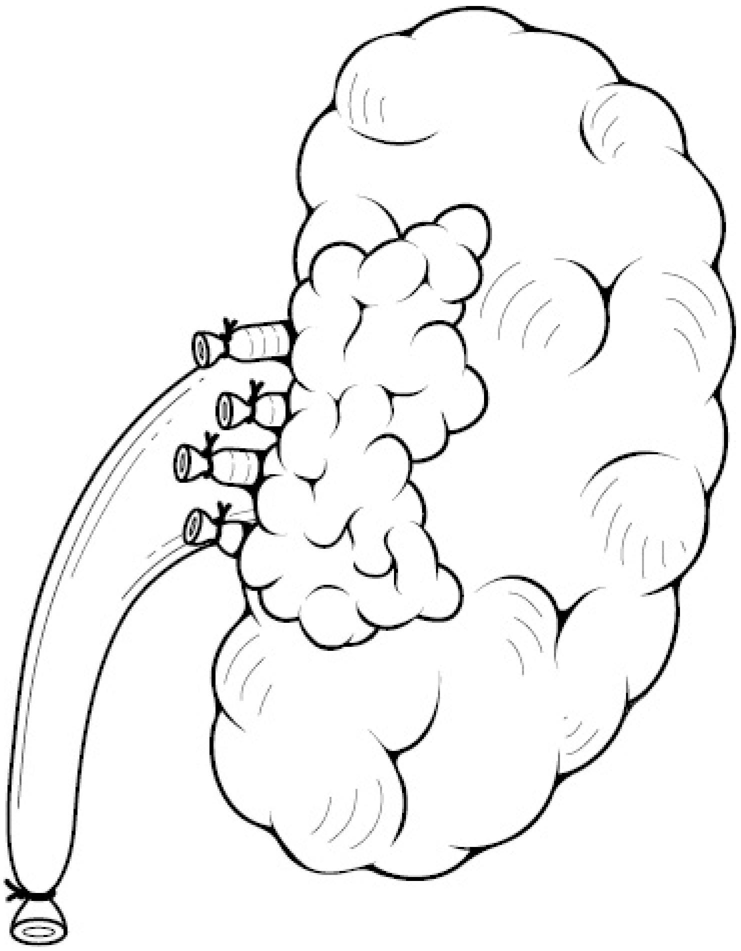
Small dysplastic kidneys might be difficult to identify immediately and care must be taken in the identification of the Gerota's fascia, as not uncommonly the peritoneum is opened instead. If this happens, the peritoneal window should be closed and the Gerota's fascia identified more posteriorly.

Once the Gerota's fascia is opened, the perinephric fat is dissected by both blunt dissection and bipolar diathermy to expose the hilar vessels and ureter (**Figure 68.4**). This step is easily accomplished by gradually pushing the kidney laterally and the perinephric fat medially, cleaning the kidney surface from the posterior to the anterior surface where renal hilum and ureter are then identified. The vessels should be individually isolated, ligated, and divided. The arterial supply should be divided first. In the case of small dysplastic or MCDK, the entire vascular pedicle can be ligated and divided together.



68.4

After the vascular pedicle has been divided, the kidney is mobilized on all surfaces and delivered from the wound (**Figure 68.5**). The attached ureter is then traced distally as far as possible (usually the level of the pelvic brim) and divided. The ureter can be ligated or left open, depending on the presence or absence of ipsilateral VUR. When a urethral catheter is placed, this is removed after 24–48 hours.

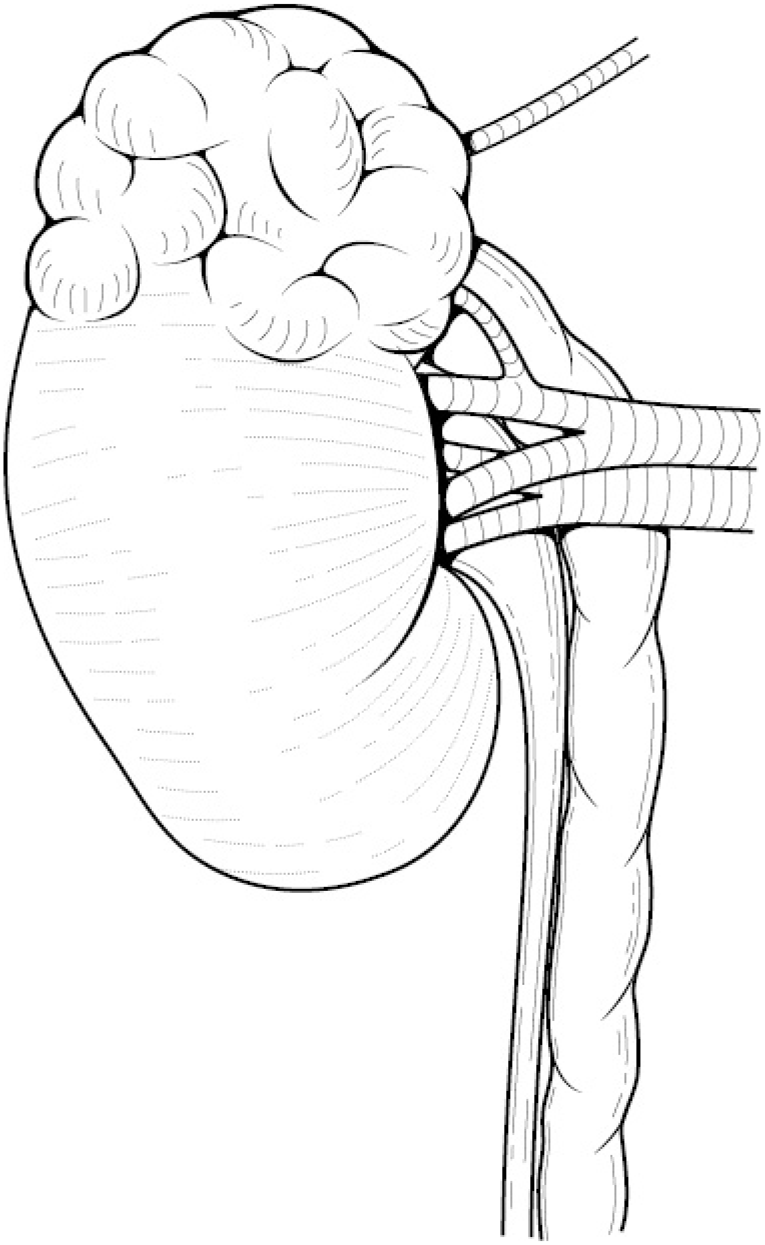


68.5

Partial nephrectomy

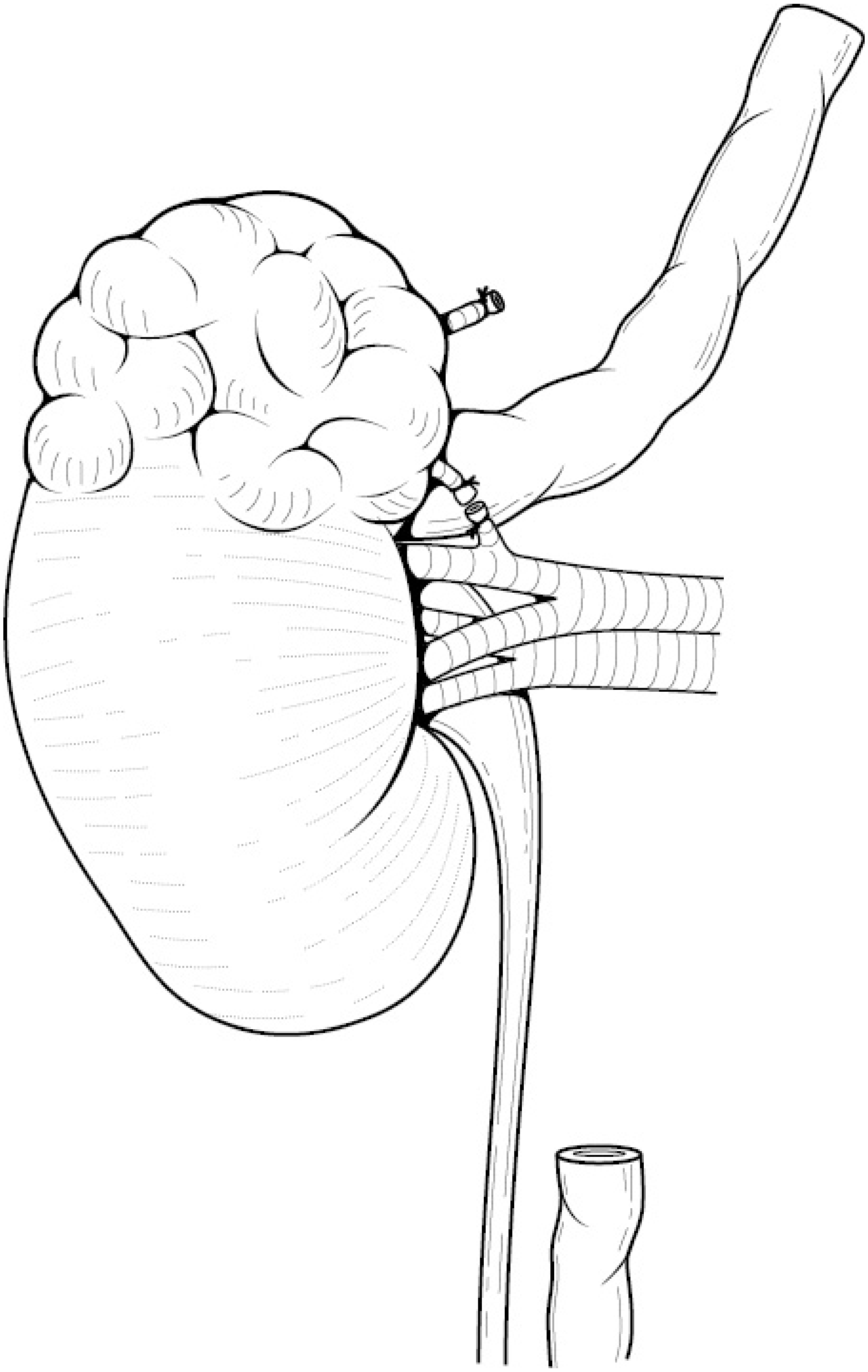
The position of the patient and the initial incision are the same for a partial nephrectomy as they are for a nephrectomy. The procedure begins as for a nephrectomy.

Once the wound has been opened and Gerota's fascia has been incised, the dissection proceeds to expose the anteromedial surface of the kidney and display the renal hilum, as for a nephrectomy. It is mandatory to visualize clearly both upper and lower moiety ureters from the outset, as this will confirm the duplex anatomy and provide a guide to the vascular supply to both renal moieties (**Figure 68.6**). The vessels supplying the affected moiety are individually identified, ligated, and divided. Often, particularly in relation to the upper renal moiety, there are short segmental vessels originating from the main vessels close to the renal hilum.



68.6

The ureter from the affected moiety is separated from the unaffected moiety ureter and divided just distal to the UPJ (**Figure 68.7**). The proximal stump of this ureter is lifted cranially, rotating it laterally to expose the posteromedial surface. There may be additional vessels supplying the affected moiety, which only become evident with this maneuver. These should be ligated and divided. Maintaining the dissection plane close to the wall of the transected ureter allows access into the renal sinus between the two collecting systems.

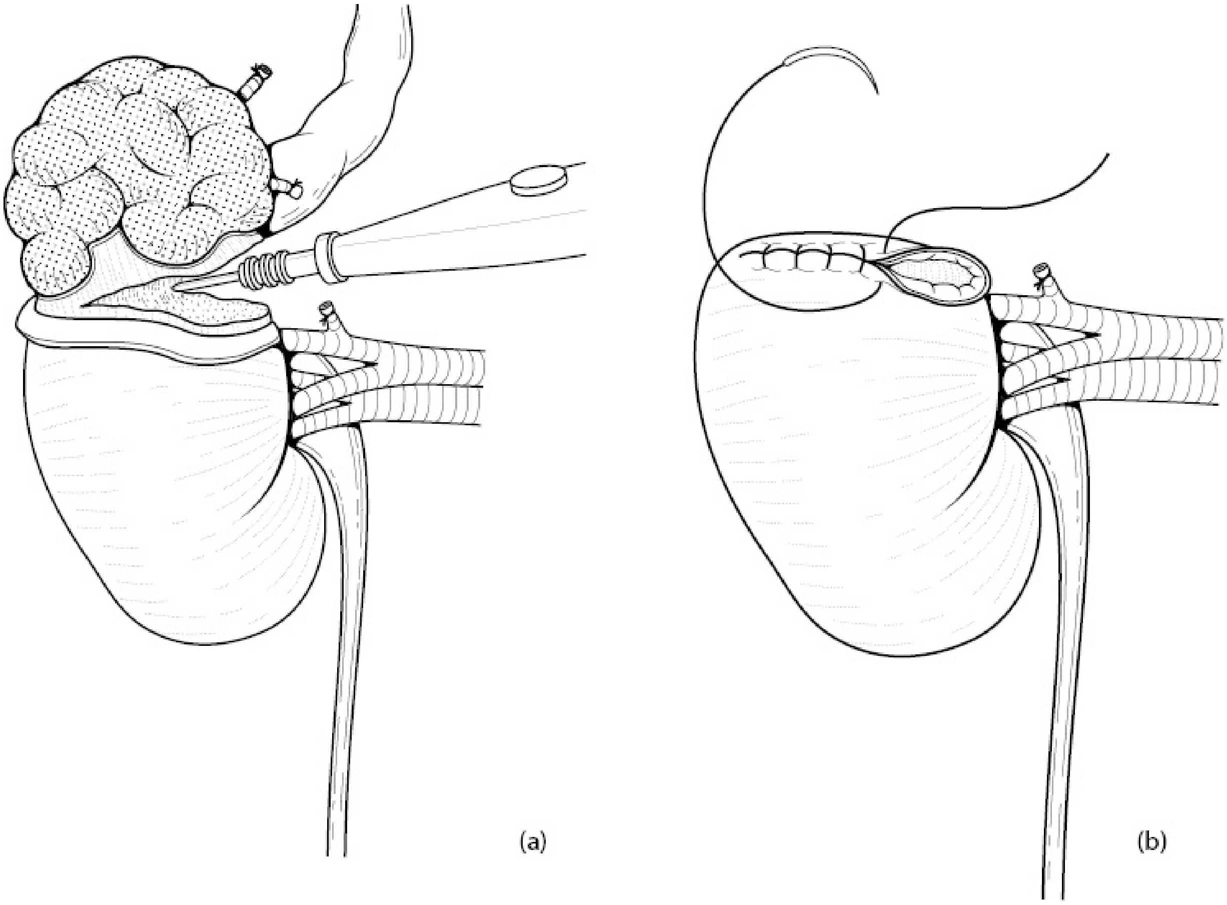


68.7

Once the vascular supply to the affected moiety has been divided, there will be purplish discoloration of the parenchyma indicating hypoperfusion.

This will serve as a guide to the line of resection through the renal parenchyma. The renal capsule is scored with monopolar diathermy at the junction between the two moieties.

The affected moiety is excised with monopolar diathermy (**Figure 68.8a**) or with a hand-held LigaSure™ (Valleylab, Boulder, CO, USA), and control of bleeding is achieved with a combination of gentle manual compression, cautery, and suture ligation. Mattress sutures of a 3/0 or 4/0 braided absorbable suture are placed to approximate the cut surfaces of the renal parenchyma (**Figure 68.8b**). Pledgets of perirenal fat or SURGICEL® (oxidized, regenerated cellulose; Ethicon Inc., Cincinnati, OH, USA) can be used to facilitate hemostasis and to prevent the suture cutting through the renal parenchyma.



68.8a,b

The distal ureteric stump should be removed as far distally as possible. Care must be taken to clearly visualize the ureter from the unaffected moiety, especially in the pelvis where they lie within a joint vascular sheath. The ureter should be ligated in all cases with documented VUR into the affected ureter.

Postoperative care

Bleeding from a slipped ligature or the cut renal surface is the earliest complication within the immediate postoperative period. Children undergoing partial nephrectomy are more at risk of developing postoperative pyrexia, and IV antibiotics may need to be given postoperatively if this occurs. Most patients can be mobilized the day after surgery and discharged from hospital once they are stable.

COMPLICATIONS

Urine leak

A retroperitoneal urinoma can occur from the reflux of urine from the distal ureteric stump or from the cut surface of the kidney following partial nephrectomy. The risk can be minimized by careful attention to the possibility of entering the collecting system during transection of the renal parenchyma. When this occurs, the defect should be closed and if necessary a double-J stent could be left *in situ*. Most urinomas will, however, resolve with the placement of a urethral catheter for at least 48–72 hours. A persistent urine leak or an infected urinoma may require the placement of a percutaneous wound drain.

Atrophy of the remnant renal moiety

This complication can occur as the result of vasospasm, thermal or traction injury to the renal pedicle. In some cases, where there is documented fever in the postoperative period, LOF can be the result of a pyelonephritic episode in the renal remnant. Prevention of this complication is the most important factor, as recognition is extremely difficult in the postoperative period. The LOF will often go unrecognized until the child attends for routine follow-up investigations.

LAPAROSCOPIC NEPHRECTOMY

HISTORY

Minimally invasive renal surgery has now become the standard and is gradually replacing open surgery. The TP approach was initially popular due to the familiarity of surgeons with laparoscopic gastrointestinal surgery. Since the description of the access technique for the retroperitoneoscopic (RP) approach, this has found favor with many surgeons. First described for end-stage renal disease patients requiring bilateral native nephrectomy, it has brought enormous advantages due to the preservation of the peritoneal integrity and the possibility to restart peritoneal dialysis immediately. Regardless of the approach utilized, the benefits in terms of a faster postoperative recovery and improved cosmesis are well recognized.

PRINCIPLES AND JUSTIFICATION

Indications

Clinical indications for laparoscopic nephrectomy/heminephrectomy are the same as for the open procedure.

Radical nephrectomy by a minimally invasive approach for renal tumors (most commonly Wilms' tumor) is permitted by SIOP guidelines for selected cases. A TP approach is required and the entire specimen needs to be retrieved through a Pfannenstiel incision.

OPERATIONS

Approach

The choice is essentially between a TP and an RP approach, with each having its own merits. The RP approach avoids colonic mobilization, the risk of injury to hollow viscera, and the potential risk of adhesion formation. However, it is believed to be more difficult to master, due to the learning curve needed for the creation of the retroperitoneal space and, once in, the reversed orientation of the kidney and hilum with the patient usually in a prone or semi-prone position. Another possible advantage of the RP approach is reduced postoperative pain due to the absence of peritoneal irritation by blood and/or urine.

Some surgeons prefer the TP approach for operations such as pyeloplasty, as it provides for a larger working space, which facilitates intracorporeal suturing. It may also be preferable in children who have previously undergone surgery to the affected kidney, in whom fibrosis and scarring may prevent the creation of a retroperitoneal working space. Inevitably, the choice of approach will also be influenced by the surgeon's experience and training.

Current evidence regarding the best approach reveals conflict between the TP and RP approaches, and is highly dependent on patient selection, center exposure, and, ultimately, the surgeon's experience. However, outcome data from a recent review have shown similar complication rates, length of surgery, and length of hospitalization between both TP and RP approaches.

In recent years, modifications to the traditional TP and RP approaches have been described, especially with regard to the number/diameter of instruments or number of incisions used to complete the procedure:

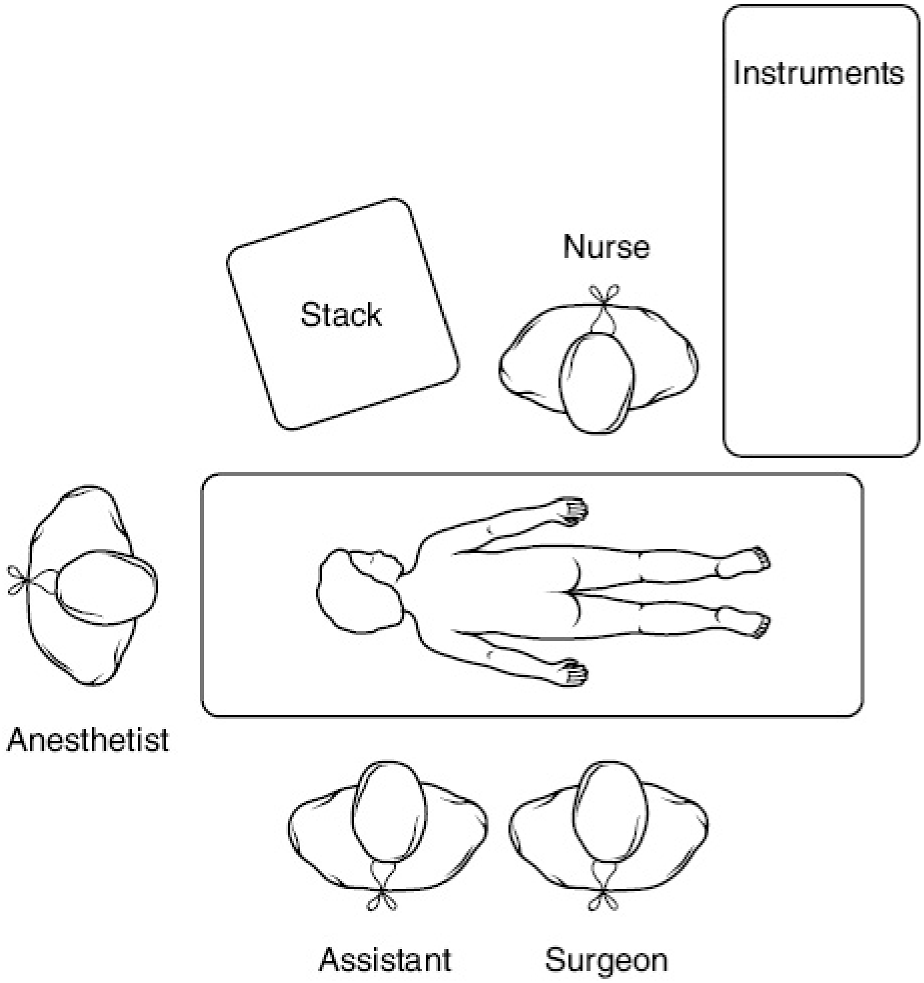
- Single-incision prone RP nephrectomy
- Laparoendoscopic single-site surgery (LESS)
- Retroperitoneal single-instrument port laparoscopic nephrectomy (SIMPL)

In this chapter, only the posterior prone RP approach will be described, as this is the senior author's technique of choice for minimally invasive nephrectomy and heminephrectomy. Of note, RP is also described through a lateral approach (with the patient in lateral decubitus position). This should allow a more extensive excision of the ureter and improved access to ectopic kidneys. However, the risk of peritoneal tears may be higher and the access to renal pedicle less direct compared to the posterior RP approach.

RP nephrectomy

ROOM SET-UP

The laparoscopic stack system should be placed on the side opposite to the affected kidney, toward the head of the table, with the screen pointing toward the pelvis. The scrub nurse should be positioned adjacent to the stack, with both the operating surgeon and assistant on the side of the affected kidney (**Figure 68.9**).

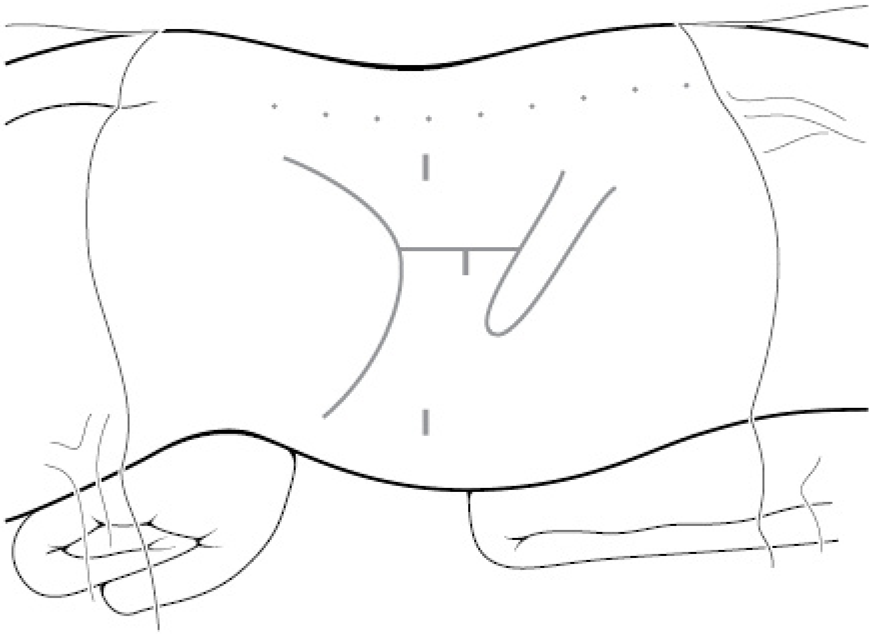


68.9

SURGICAL ACCESS

The patient is positioned fully prone under general anesthesia. The exposed dorsal and lateral aspects of the trunk are prepared and draped in a sterile manner. Topographic landmarks and anticipated port sites are marked as shown.

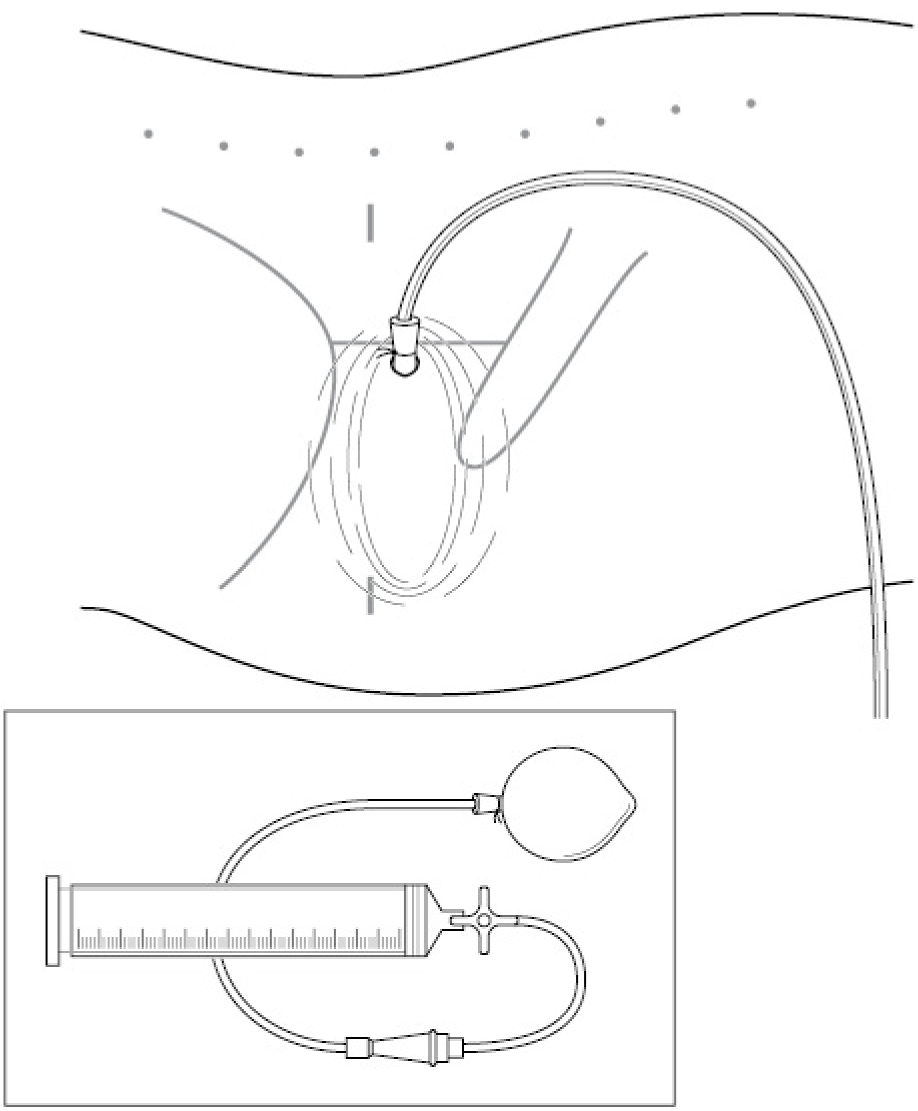
A 5 mm transverse incision is made midway between the iliac crest and the tip of the twelfth rib, just lateral to the outer border of the sacrospinalis muscle (**Figure 68.10**). Through this incision, a small area of the retroperitoneum is dissected bluntly with artery forceps to allow insertion of the retroperitoneal dissecting device.



68.10

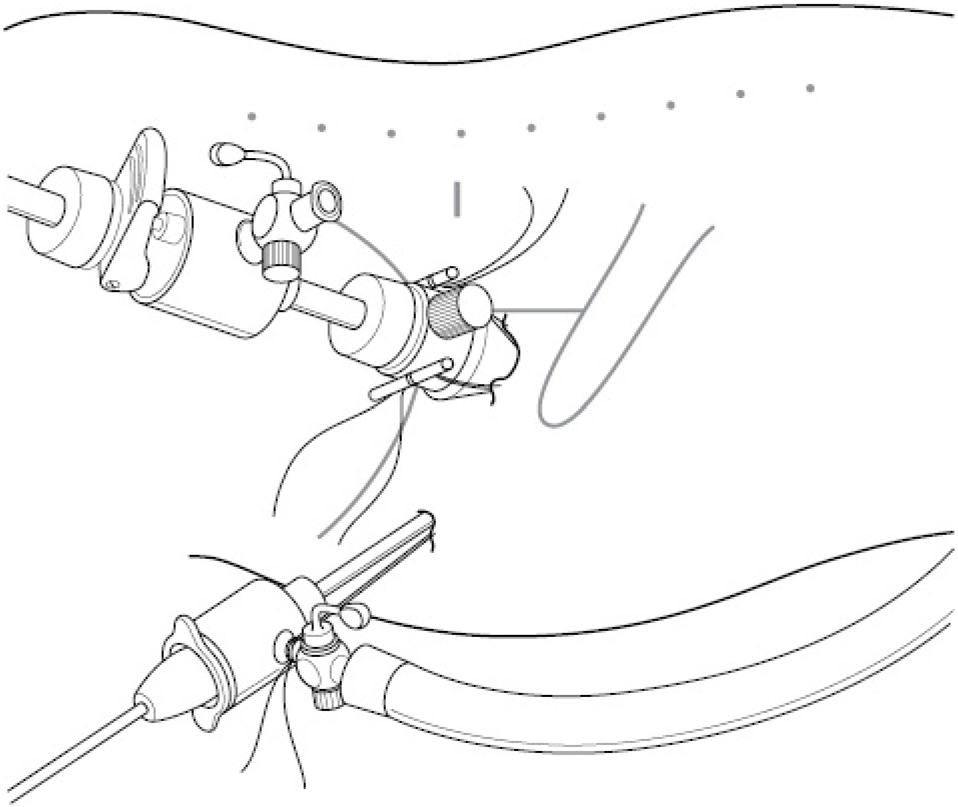
PROCEDURE

The retroperitoneal space is developed outside Gerota's fascia by a technique described by Gill et al. (1996). A dissecting balloon is made by securing the finger of a sterile surgical glove to the end of a 12 Fr Jacques catheter with a silk tie. The catheter is connected to a three-way tap and a 50 mL Luerlock syringe. Depending on the size of the patient, 100–250 mL of air is injected slowly to develop the retroperitoneal space. The balloon is left inflated for 1 minute to promote hemostasis, and then deflated and withdrawn (**Figure 68.11**).



68.11

A 6 mm Hasson cannula is inserted into the port site, followed by insufflation of the retroperitoneum with carbon dioxide to a pressure of 10–12 mmHg. The Hasson port is secured by a suture to the skin. A 5 mm instrument port is placed under direct vision below the tip of the eleventh rib and above the iliac crest (**Figure 68.12**). A 30-degree 5 mm laparoscope is recommended for all RP surgery, as it can be rotated along its longitudinal axis to alter and maximize the viewpoint for the surgeon.

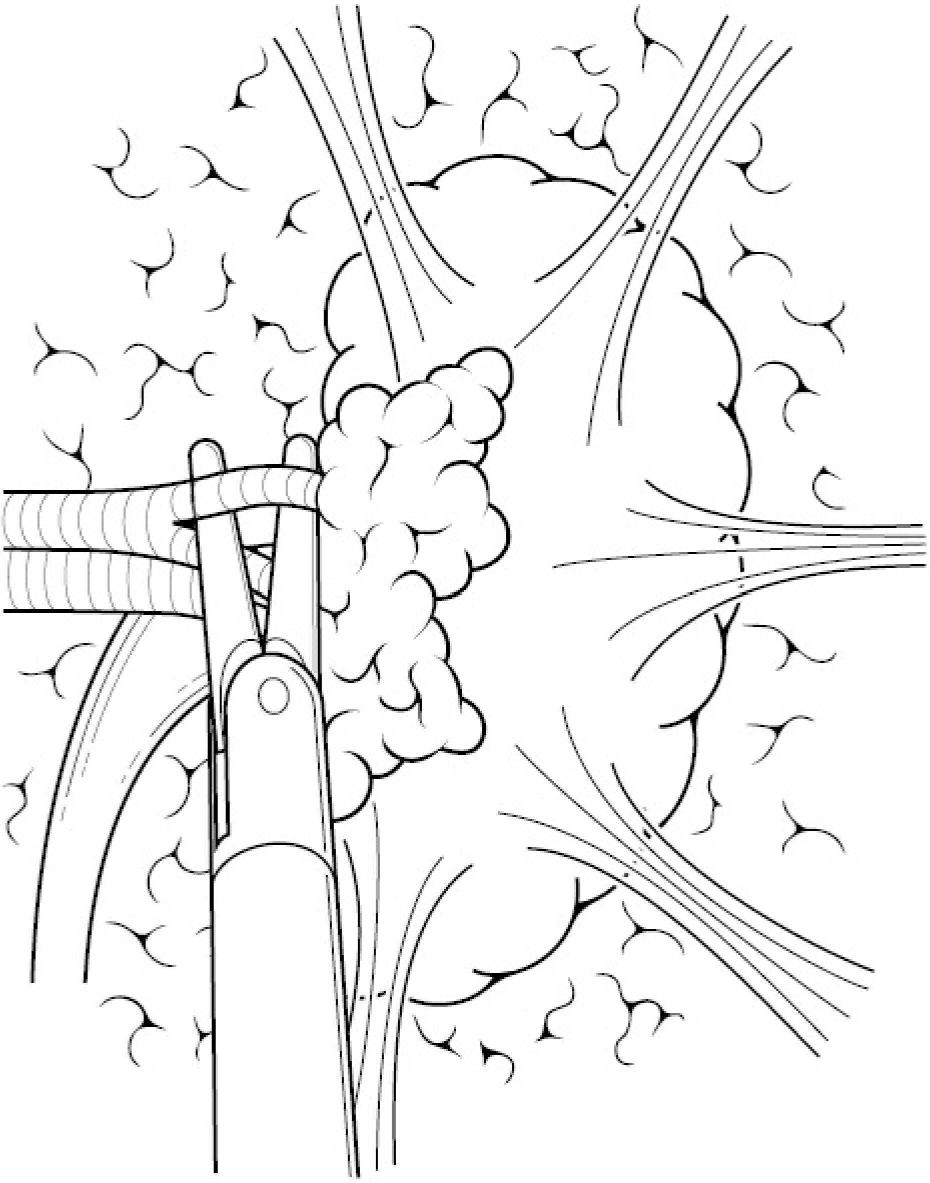


68.12

Gerota's fascia is incised longitudinally adjacent to the posterior abdominal wall using scissors. A cruciform incision can also be chosen to open the Gerota's fascia. Bleeding points are meticulously controlled with monopolar diathermy. The loose areolar adventitial tissue can then be visualized through the window in Gerota's fascia. This tissue is dissected in a blunt manner to create a large perinephric working space, thereby exposing the posterolateral surface of the kidney.

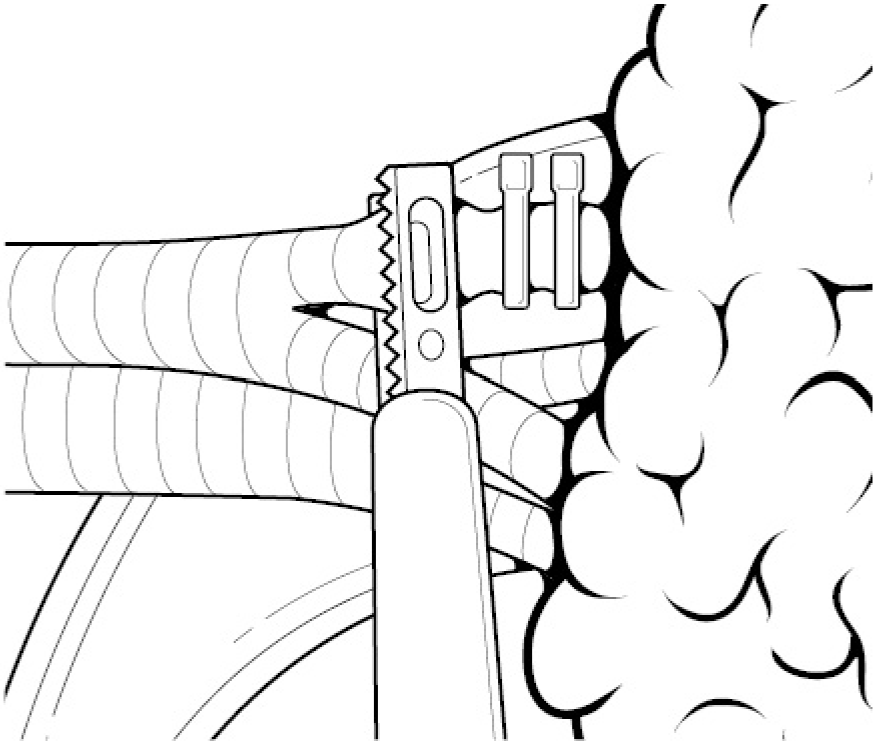
In the authors' experience, the entire operation can be performed safely through a single instrument port. However, for surgeons at the beginning of the learning curve, it is recommended that a second instrument should be placed to firstly achieve confidence with the approach. This second instrument provides countertraction for dissection and easier application of hemoclips.

Dissection is started at the apex and continued along the medial aspect of the kidney, pushing it laterally and anteriorly to expose the posteromedial surface further, in particular, the renal hilum (**Figure 68.13**). The lateral and inferior attachments are maintained intact to facilitate exposure of the renal pedicle by the traction of the kidney by these attachments.



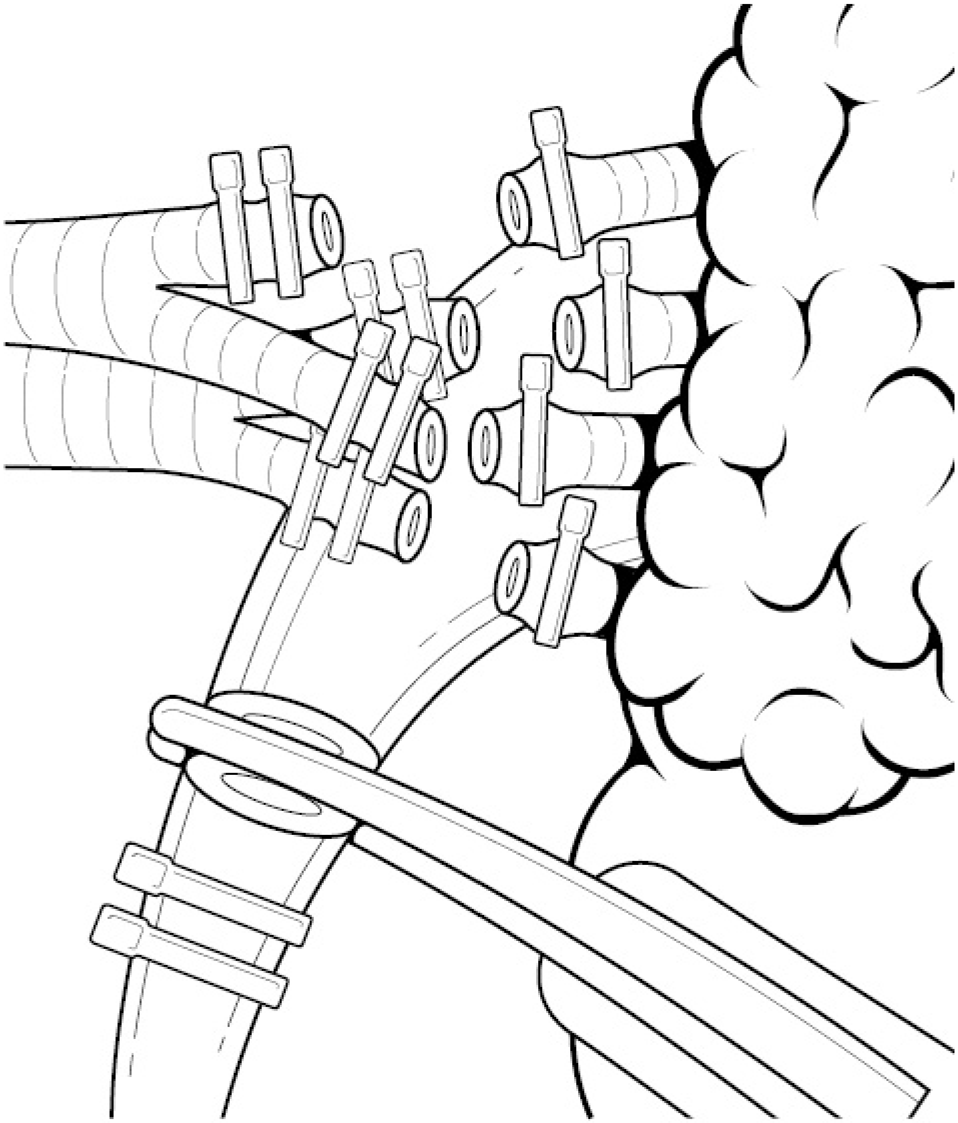
68.13

After adequate mobilization of the renal pedicle, the vessels are divided between hemoclips or with a LigaSure™ (Valleylab, Boulder, CO, USA) when the vessels are less than 6 mm in diameter (**Figure 68.14**). A minimum of three clips should be applied on all vessels, with at least two clips remaining on the proximal stump of the divided vessel. Care must be taken to identify and divide every possible vessel, particularly in the case of MCDKs, in which anomalous vessels are frequently found.



68.14

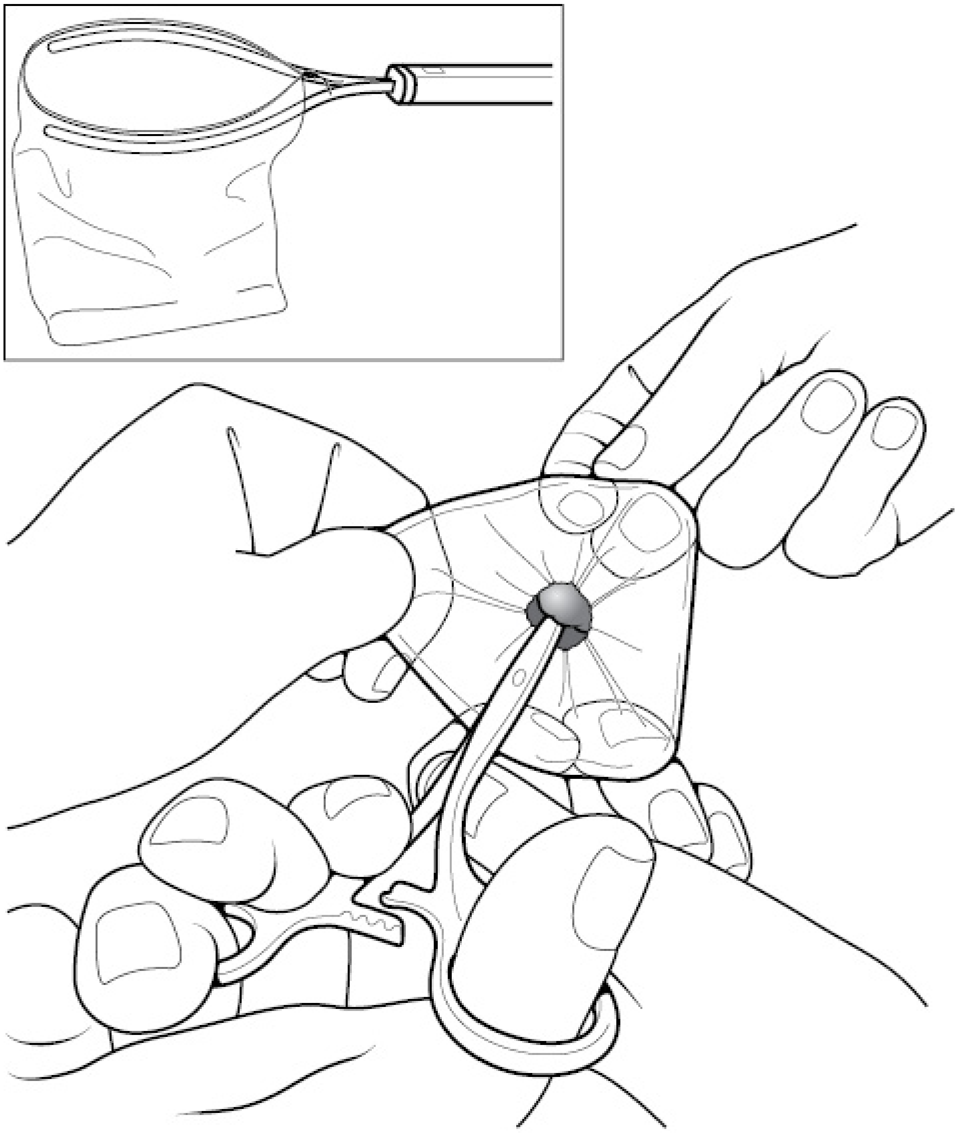
The ureter is dissected distally as far as required and divided/ligated (**Figure 68.15**). If a near-complete ureterectomy is intended, such as in cases with reflux-associated nephropathy, the RP approach will allow access into the pelvis to just below the level of the pelvic brim. In all cases of documented ipsilateral reflux, it is the author's practice to ligate the ureter with a 3/0 Vicryl™ endoloop suture. If this were not possible, an alternative approach would be to leave the ureteric stump open and to drain the bladder with an in-dwelling urethral catheter for 48–72 hours. In the absence of ipsilateral VUR, the ureter can be safely left open without postoperative bladder drainage.



68.15

The remaining attachments of the kidney are divided using a combination of blunt dissection, monopolar diathermy, and/or LigaSure™ (Valleylab, Boulder, CO, USA). In the case of a large MCDK, complete intracorporeal mobilization can be technically difficult, time-consuming, and at risk of creating a tear in the closely attached peritoneum. In such cases, after all vessels have been divided and the cysts decompressed by percutaneous punctures, the kidney can be withdrawn via the camera port incision and the remainder of the dissection completed in an extracorporeal manner.

The specimen can be extracted directly through the camera port incision in the case of a MCDK, grossly hydronephrotic kidney, or a small dysplastic kidney. Larger specimens are extracted with the use of a 10 mm endobag specimen retrieval device (**Figure 68.16**). The specimen is entrapped within the endobag and removed piecemeal with the use of sponge-holding forceps. The wound is closed in layers. Usually no percutaneous drain is needed.

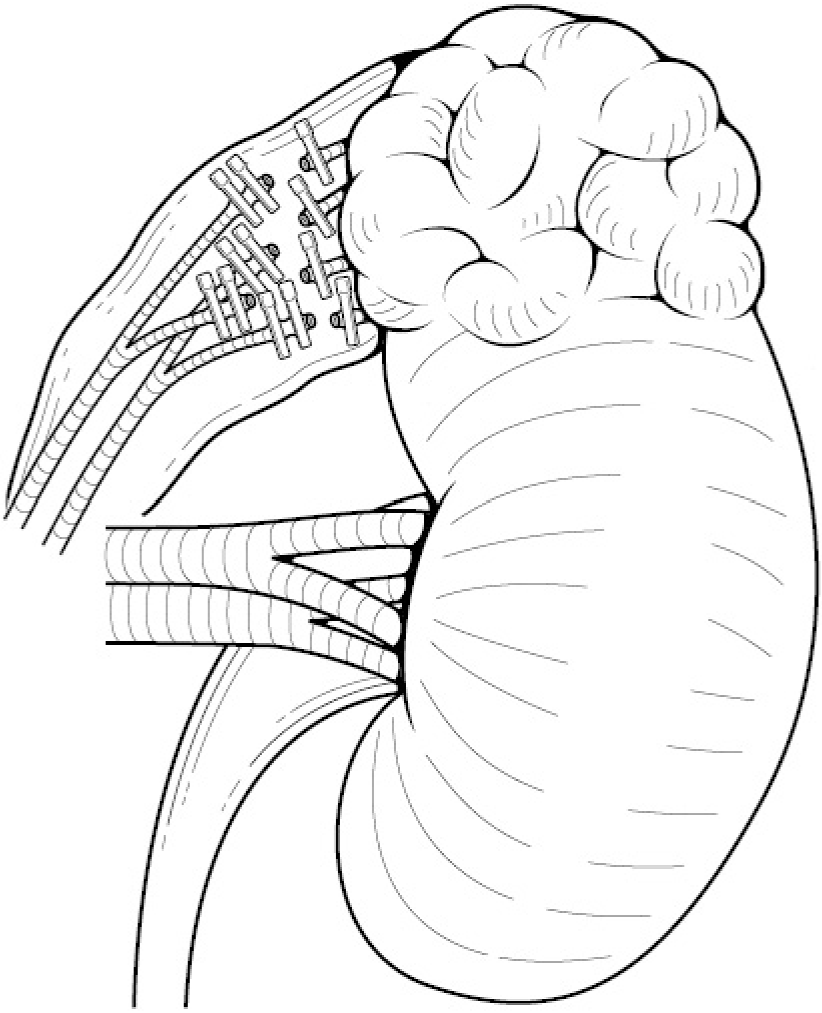


68.16

RP heminephrectomy

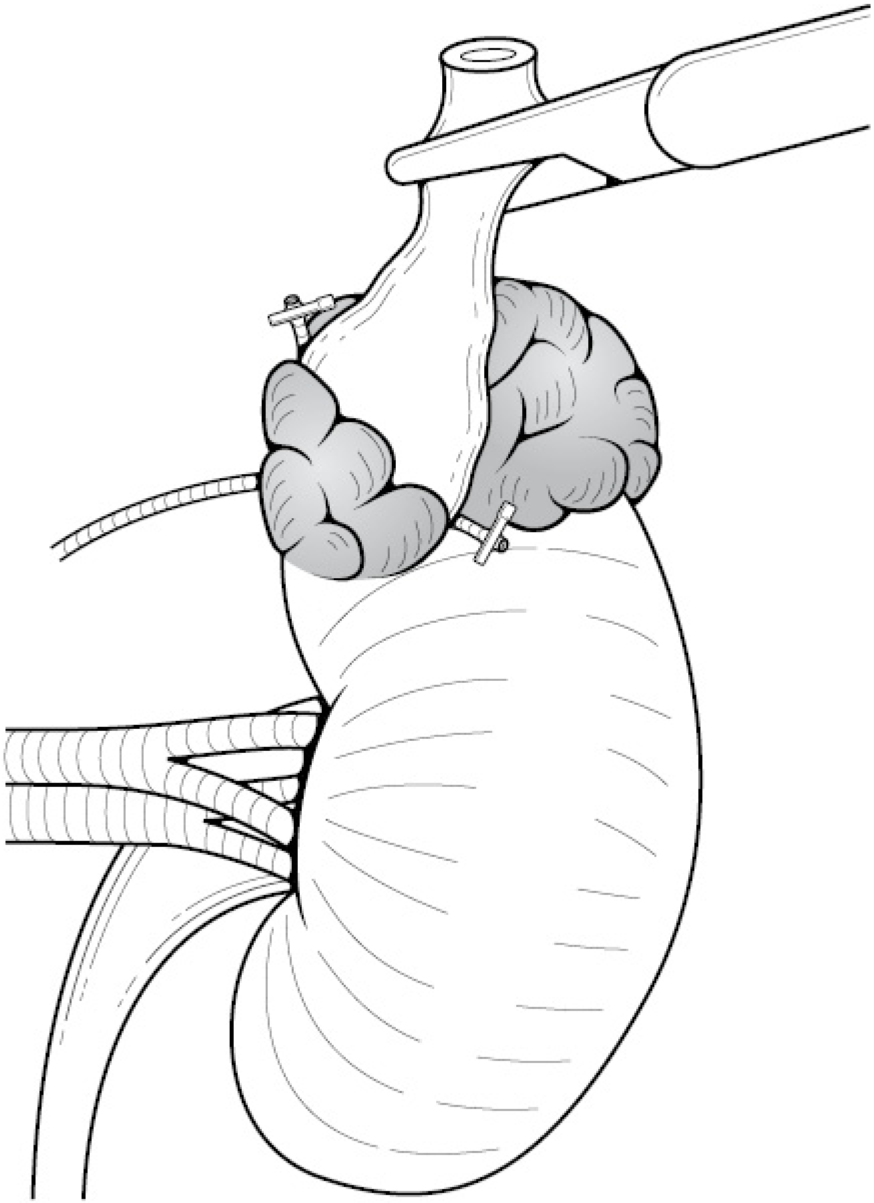
Once the retroperitoneal space has been created and Gerota's fascia has been incised, the dissection proceeds to expose the posterolateral surface of the kidney and display renal hilum as for a nephrectomy. It is mandatory to visualize clearly both upper and lower moiety ureters from the outset, as this will confirm the anatomy and provide a guide to the vascular supply to both renal moieties.

The vessels supplying the affected moiety are selectively identified and divided between clips or with an energy device (**Figure 68.17**). In some cases, the polar vessels will be clearly evident, whereas in other cases there will be short segmental vessels originating from the main vessels close to the renal hilum. The latter scenario is seen more frequently when the affected renal moiety is small and dysplastic.



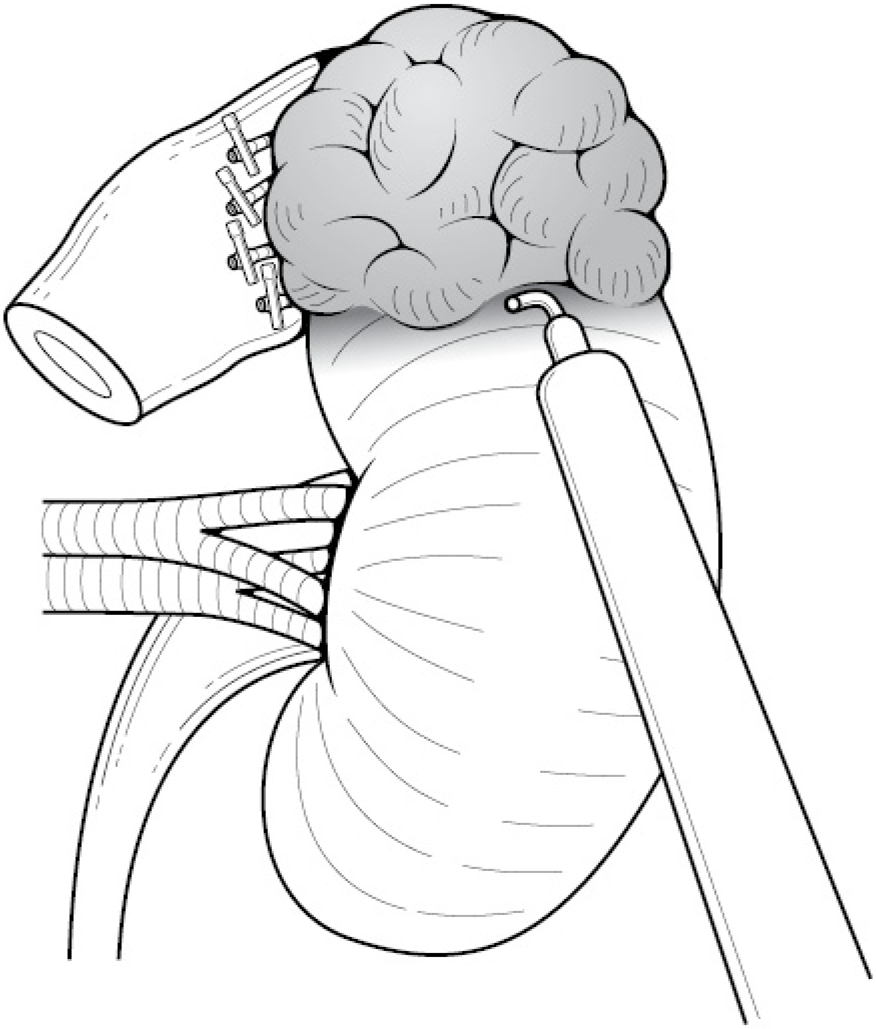
68.17

The ureter from the affected moiety is separated from the non-diseased ureter and divided just distal to the UPJ. It is important to handle the non-diseased ureter as little as possible and, if needed, very carefully, preserving its adventitia, as the ureteric vasculature is particularly delicate. The proximal stump of the affected ureter is used to lift the kidney, rotating it laterally to expose the anteromedial surface (**Figure 68.18**). This will allow the best visualization of the vasculature as there may be additional vessels supplying the affected moiety, which only become evident with this maneuver.



68.18

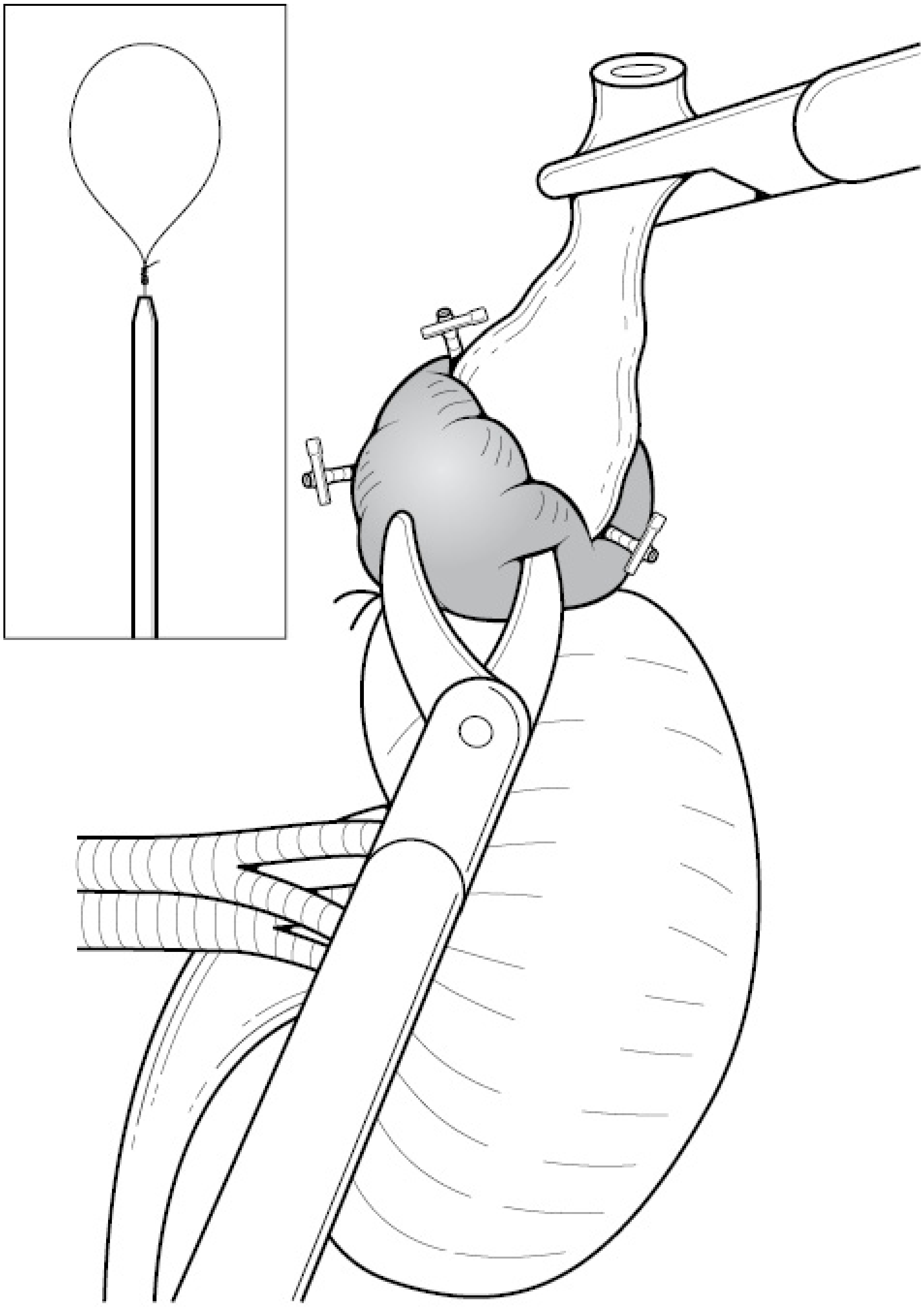
Once all the vessels to the affected moiety have been secured, there will be a clear blanching of the parenchyma, indicating hypoperfusion. This will provide a guide to the portion of renal parenchyma to resect. The renal capsule is scored with monopolar diathermy at the junction between the two moieties (**Figure 68.19**).



68.19

The affected moiety, now hypoperfused, can be removed. This can be achieved with an endoloop (e.g. ENDOLOOP®, Ethicon Inc., Cincinnati, OH, USA) or energy device.

A 3/0 Vicryl™ endoloop suture is placed over the affected moiety, using the proximal end of the divided ureter as countertraction to facilitate this sometimes difficult step (**Figure 68.20**). The ligature is firmly tightened at the junction between the renal moieties, providing secure hemostasis and minimizing the risk of urine leak. The parenchyma is transected with hook scissors 5–10 mm distal to the ligature. The cut surface is carefully inspected to detect any remaining arterial bleeding points, which are rare with the endoloop technique, but would need to be secure with diathermy or a further endoloop suture. In the author's center experience the use of endoloop has resulted in more cyst formation at follow-up compared with energy devices.



68.20

An energy device (e.g. LigaSure™ [Valleylab, Boulder, CO, USA]; Harmonic scalpel™ [Ethicon Inc., Cincinnati, OH, USA]) is used to progressively follow the scored renal capsule, checking carefully for bleeding after each advancement.

The distal ureteric stump should be removed as far distally as possible. Care must be taken to visualize clearly the ureter from the unaffected moiety, especially in the pelvis, where both ureters lie within a common sheath. The ureter should be ligated in all cases with documented VUR into it.

The specimen can be extracted directly through the camera port incision in the majority of cases, as usually those are small dysplastic upper moieties. Larger specimens are extracted with the use of a 5–10 mm endopouch specimen retrieval device. The wound is closed in layers. The use of percutaneous drainage is usually unnecessary.

POSTOPERATIVE CARE

As before, the immediate complication to be wary of is bleeding with later urine leak.

Mean reported length of hospitalization is 2–3 days for both TP and RP total nephrectomy. However, some experienced centers discharge the patient on day one postoperatively.

COMPLICATIONS

Peritoneal tear

The posterior prone approach minimizes the risk of a peritoneal tear as compared to other approaches for RP surgery, when the balloon is too small for the size of the patient, in adolescence, and in children on peritoneal dialysis.

With the posterior prone approach the risk of a peritoneal tear is minimal, but can occur when performing pre-transplant nephrectomy for nephrotic syndrome, where the kidneys are often large or when a child is on peritoneal dialysis, where the peritoneum is very thin and delicate. Peritoneal tears can also occur when the dissecting balloon is inflated too rapidly. The impact of a peritoneal tear on the working space is very unpredictable but generally reduces the working space. If the tear is very small, making the tear larger can, at times, allow equilibration of the gas pressures with an increase in the working space. Another option would be to repair the tear with sutures.

Balloon rupture

Rupture of the dissecting balloon can occur when the balloon is inflated too rapidly, with overinflation of the balloon, or when excessive external pressure is applied over the balloon. When it occurs, the ruptured balloon must be carefully examined for lost fragments, which should be sought and removed from the retroperitoneal space.

Intraoperative bleeding

Intraoperative bleeding is most likely to be due to slipping of hemoclips from a renal vein or to inadvertent damage to a renal vein or vena cava by a laparoscopic instrument. In most cases, hemorrhage can be controlled by the prompt application of hemoclips to the affected vessel or side wall of the vena cava. Uncontrollable hemorrhage will require conversion to an open approach to ligate or oversew the bleeding vessel.

Urine leak

A retroperitoneal urinoma can occur from the reflux of urine from the distal ureteric stump or from the cut surface of the kidney following heminephrectomy. The risk can be kept to a minimum by the use of the energy devices on the renal parenchyma and by endoloop ligation of refluxing ureters as opposed to the use of hemoclips or an energy device to seal the ureter. Most urinomas will resolve with the placement of a urethral catheter for at least 48–72 hours. A persistent urine leak or an infected urinoma may require the placement of a percutaneous wound drain. Theoretically, with the RP approach the urinoma should be confined and resolve more rapidly compared with TP surgery, where the peritoneal cavity could allow the leak to drain for longer.

Cyst formation

The development of a cyst at the parenchymal resection margin after a laparoscopic heminephrectomy has a reported incidence of up to 50%. In the authors' experience, the overall incidence of polar cyst formation following 111 TP and RP heminephrectomies was 39% (43/111), with the vast majority of cysts following lower-pole (80%) compared to upper-pole surgery (20%) and with the use of endoloop technique (70%) compared to transection of the renal parenchyma with an energy device. No cysts required surgical reintervention.

Loss of function of remaining moiety after RP heminephrectomy

LOF after RP upper-pole heminephrectomy should not be expected. Partial LOF (mean LOF 10%) has been reported by recent series with an incidence around 17%. In the senior author's experience, significant LOF of the remaining moiety following retroperitoneal heminephrectomy occurred in around 5% of cases. Although no specific causes have been associated with this significant LOF, young age at surgery (<1 year), upper-pole heminephrectomy, and presence of postoperative complications may increase this risk.

VIDEOS

OPS_8e_video_68.1 Left lower pole heminephrectomy
OPS_8e_video_68.2 Retro right nephrectomy
OPS_8e_video_68.3 Right upper pole heminephrectomy

FURTHER READING

El-Ghoneimi A, Abou-Hashim H, Bonnard A, et al. Retroperitoneal laparoscopic nephrectomy in children: At last the gold standard? *J Pediatr Urol* 2006; 2: 357–63.

Esposito C, Escolino M, Troncoso Solar B, et al. Diagnosis and long-term outcome of renal cysts after laparoscopic partial nephrectomy in children. *Br J Urol Int* 2017; 119: 761–6.

Featherstone NC, De Win G, Undre S, Cherian A. Single incision prone retroperitoneoscopic paediatric nephrectomy. *J Pediatr Urol* 2015; 11: 283–4.

Gill IS, Grune MT, Munch LC. Access technique for retroperitoneoscopy. *J Urol* 1996; 156: 1120–4.

Jayram G, Roberts J, Hernandez A, et al. Outcomes and fate of the remnant moiety following laparoscopic heminephrectomy for duplex kidney: A multicenter review. *J Pediatr Urol* 2011; 7: 272–5.

Kim C, McKay K, Docimo SG. Laparoscopic nephrectomy in children: Systematic review of transperitoneal and retroperitoneal approaches. *Urology* 2009; 73: 280–4.

69

Vesicoureteric reflux: Open and minimally invasive treatment

AURELIEN SCALABRE, DELPHINE DEMÈDE, AND PIERRE MOURIQUAND

HISTORY

Pozzi first described the concept of vesicoureteric reflux (VUR) at the end of the 19th century having accidentally transected a ureter during pelvic surgery! The antireflux mechanism of the vesicoureteric junction (VUJ) was reported by Sampson in 1903, and in 1923 Graves and Davidoff stated that reflux is not normal. VUR and its renal consequences (chronic atrophic pyelonephritis) remained poorly understood until 1960. Children with recurrent urinary tract infections (UTIs) tended to be treated by surgical procedures to the bladder neck and urethra. Nephrectomy for recurrent unilateral symptomatic pyelonephritis was not uncommon. In 1960, Hodson and Edwards clarified the relationship between VUR and renal damage and stressed the importance of pyelotubular backflow of urine into certain papillae. In 1973, Bailey introduced the term "reflux nephropathy," and later Ransley and Risdon published an essential study on papillary morphology. The concept of early damage of renal parenchyma ("big bang") at the first UTI in infants with VUR and the potential danger of intrarenal reflux of infected urine was proposed by Ransley and Risdon, and Smellie et al. in 1975. Surgical correction of VUR was widely practiced until it was documented that, in many cases, the reflux resolved spontaneously with growth of the child, and the only treatment required was prophylaxis against UTIs. Antenatal detection of urinary dilatation and suspicion of reflux provided an opportunity to start prophylaxis soon after birth.

PRINCIPLES AND JUSTIFICATION

Definition

VUR may be defined as a "permanent or intermittent intrusion of bladder urine into the upper urinary tract due to a defective ureterovesical [vesicoureteric] junction."

The defect in the VUJ may be a primary disorder or may arise secondary to bladder dysfunction (neuropathic bladder, dyssynergic bladder) or bladder outlet obstruction (e.g. posterior urethral valve). The refluxing urine can fill the upper excretory system (ureters and renal pelvis) between and/or during micturition, and can sometimes penetrate into the renal substance (intrarenal reflux). The volume of refluxing urine can vary in the same patient at different times. Attempts to classify the degree of reflux are therefore of limited practical interest, as reflux may change in intensity in individual patients.

Pathophysiology

The pathophysiology of VUR remains unclear, but there is a general consensus that intrarenal reflux of infected urine can cause renal damage (reflux nephropathy). It is also likely that other disorders, such as immune reactions, associated renal dysplasia, and change of urine biochemistry, can also be responsible for renal deterioration. The difficulty is that there is no investigation able to distinguish acquired or congenital renal lesions.

There are essentially two different types of clinical presentation of VUR.

- *Primary malformative VUR*, i.e. major VUR associated with dilated upper urinary tracts on prenatal and postnatal ultrasound (US) scans, abnormal dimercaptosuccinic acid (DMSA) scan in >50% of cases, and a low spontaneous regression rate. This type occurs mainly in boys (90%) and is due to a congenital malposition of the emergence of the ureteric bud (ureteral bud theory of Mackie and Stephens) resulting in a morphological abnormality at the level of the VUJ.

- *Secondary VUR,* i.e. secondary to lower urinary tract dysfunction. Abnormal bladder urodynamics are more common than in primary reflux, do not cause upper tract dilatation, and are not detectable prenatally. Secondary VUR is not associated with an abnormal DMSA scan, and usually occurs in girls with poor bladder and bowel function. The resolution rate is around 80% with bladder training. Radical treatment is seldom required.

Indications

There is failure to reach a consensus on the precise indications for surgery, and it can be difficult to select the ideal treatment for each patient. However, during the last decade several studies have shown the importance of the couple "kidney–bladder" and it is now accepted by many that VUR with normal kidney DMSA at an early stage of life will resolve spontaneously with maturation of the bladder. It is therefore logical that children with recurrent urinary tract infections (RUTIs) should undergo initial evaluation of their kidneys and only those children with abnormal DMSA would require cystography ("top-down approach"). A common policy in those with VUR is to prescribe prophylactic antibiotic treatment for prolonged periods (12–36 months) in the expectation of spontaneous maturation of the VUJ and a resolution of VUR. The long-term side effects of prophylactic treatment are unknown, and there are reservations regarding its effectiveness in preventing infection. If the child suffers from breakthrough infections or if there are signs of continued deterioration of the renal substance, more radical treatment should be considered.

Surgery consists of reimplanting the ureters into the bladder; endoscopic treatment consists of injecting a substance behind the intramural ureter to create an effective posterior backing, which helps to restore the antireflux mechanisms. The aim of these modalities of treatment is to stop the reflux. This does not necessarily mean that the progression of renal damage is halted or that further UTIs are prevented. In cases where VUR has caused severe damage to the kidney (relative function <10%), a nephroureterectomy may be the best option. The choice between surgical, laparoscopic, and endoscopic correction of reflux is an individual matter, each technique having its own protagonists. The best result with endoscopic treatment is with low-grade refluxes, where spontaneous resolution is high and where underlying bladder dysfunction is common. Although simple and less invasive, the place of endoscopic treatment is therefore unclear and one should remain cautious about the current trend to extend its indications (see Table 69.1).

Table 69.1 **Reflux symptom (bladder dysfunction) and reflux disease**

	Symptom	**Disease**
Presentation	Recurrent urinary tract infections	Prenatal diagnosis
Ultrasound	Normal	Dilatation
Cystography	Grades I–III	Grades III–V
DMSA scan	Normal	Abnormal
Resolution	>80%	<50%
Etiology	Bladder dysfunction	Abnormal VUJ + renal dysplasia
Treatment	Training	Surgery

PREOPERATIVE ASSESSMENT

Several investigations are commonly performed:

- *Cystography:* Micturating cystography is the standard method to identify and grade VUR. Its indications have certainly decreased with the "top-down approach," but it remains mandatory prior to surgery.
  - Direct contrast micturating cystography with bladder catheterization is the most common.
  - Indirect radioisotope cystography is less reliable, but may be used in follow-up. Direct isotopic micturating cystography has the advantage of a considerably reduced radiation dose, but it is not widely available.
- *DMSA or 99mTc-labeled mercaptoacetyl triglycine (MAG3) renal scanning:* This assesses the presence of renal scars and provides a measure of the relative function of each individual kidney.

- *Urodynamic study:* This is important for detecting an underlying bladder dysfunction.
- *Ultrasonography:* Although a poor investigation to detect reflux, this can be useful for assessing the size, shape, and echogenicity of each kidney, the degree of dilatation of the ureter, and bladder wall thickness.
- *Cystoscopy:* Evaluation of the shape of the ureteric orifice or the length of the submucosal portion of the ureter is often subjective and will rarely alter the therapeutic decision. It is, however, recommended if the VUR is associated with a contralateral or ipsilateral ureterocele or if the reflux has occurred secondary to bladder dysfunction or bladder outlet obstruction.

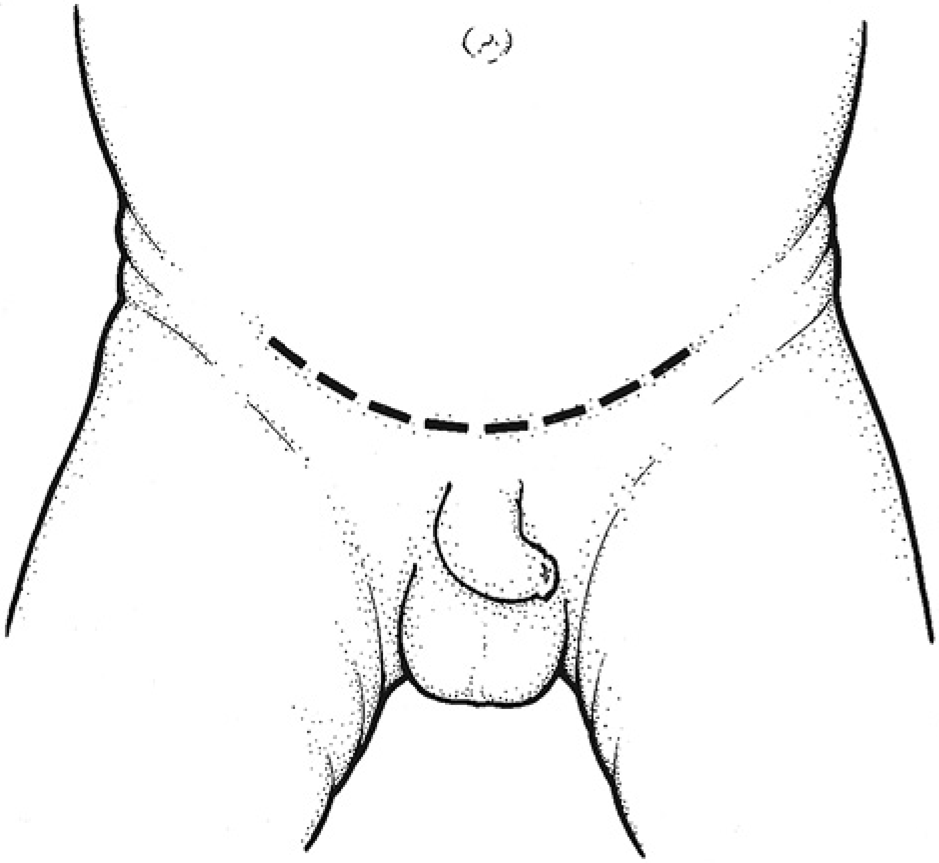
OPEN SURGERY

Transhiatal reimplantation of the ureter (typically, Cohen's or Glenn–Anderson procedure), suprahiatal reimplantation of the ureter, and extravesical reimplantation are the three main surgical options. The aim is to mobilize the distal segment of the ureter(s) (transmural ureter) and place it under a tunnel of bladder mucosa in order to restore the flap valve mechanism.

OPERATIONS

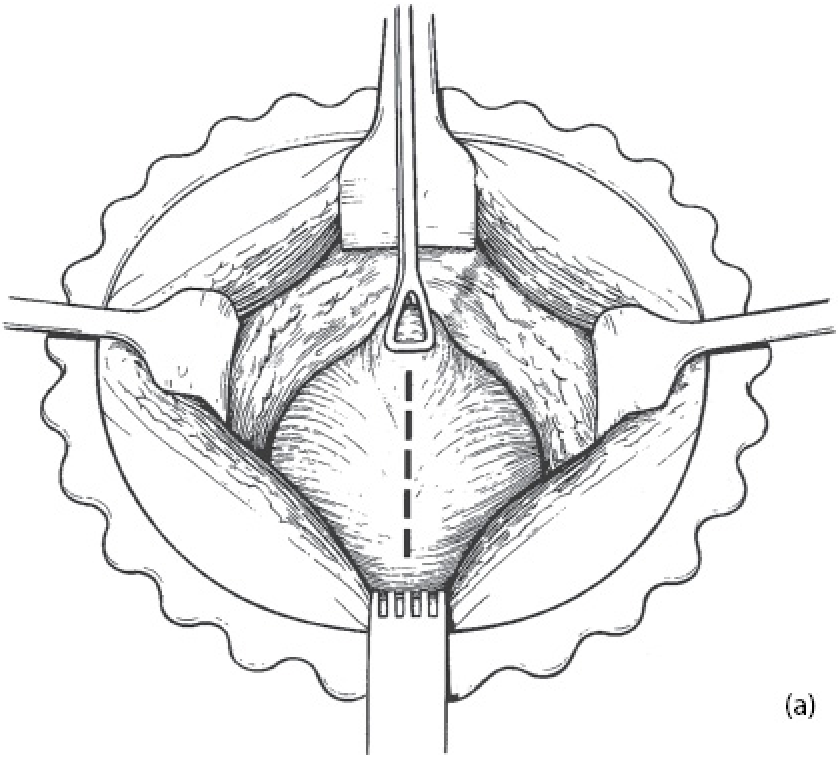
Transhiatal reimplantation of the ureter: Cohen's procedure

A transverse suprapubic incision is made 2 cm above the pubic symphysis, in the low abdominal crease (**Figure 69.1**). The subcutaneous tissues are incised, exposing the rectus sheath, which is opened vertically in the midline. Both recti are separated and the peritoneum is gently pushed upward, superiorly, providing good exposure of the bladder.

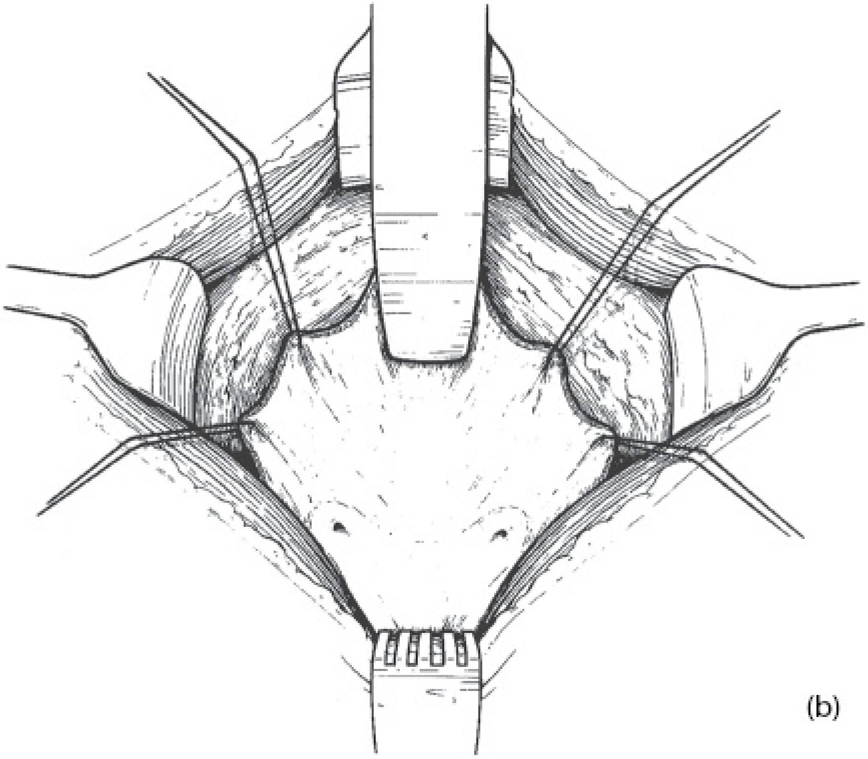


69.1

A Denis Browne retractor is inserted. The lateral blades retract the recti and upper and lower blades retract the skin and subcutaneous tissues (**Figure 69.2a**). The anterior wall of the bladder is incised vertically and two stay sutures suspend each edge of the vesicotomy (**Figure 69.2b**). Swabs are put inside the bladder and retracted upward with a Deaver retractor in order to expose the trigone. A 3/0 or 4/0 absorbable suture is placed at the lowest point of the vesicotomy to prevent splitting of the incision downward into the urethra.



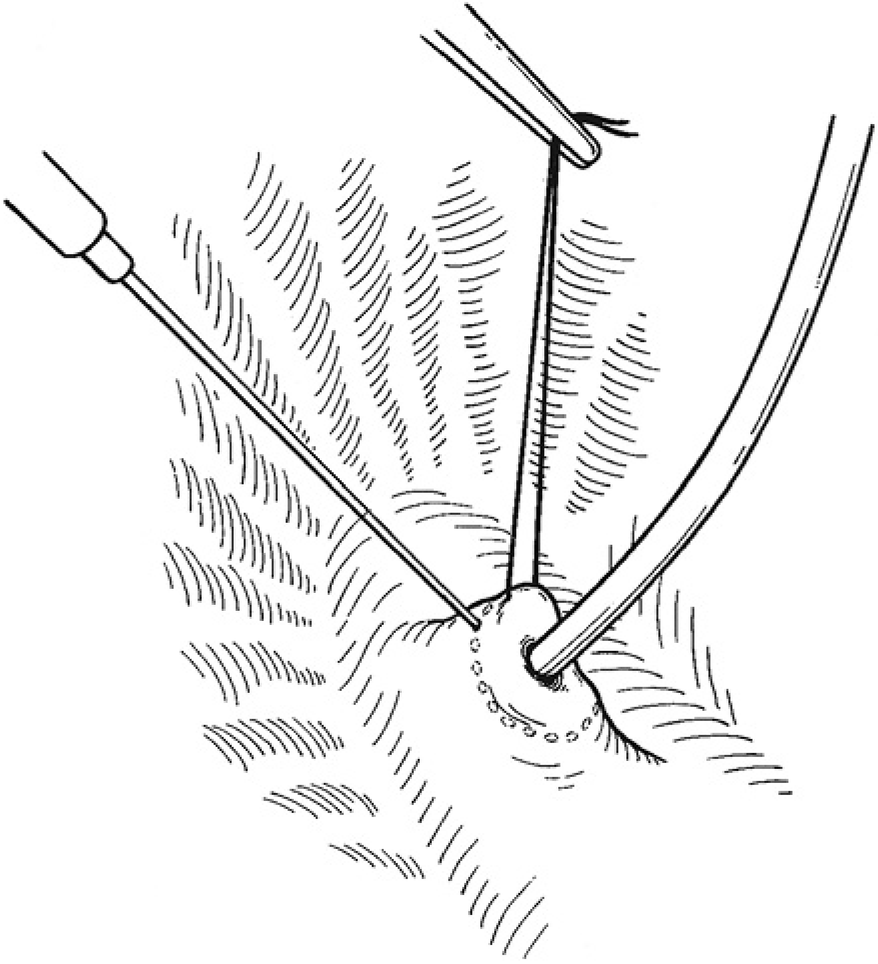
(a)



(b)

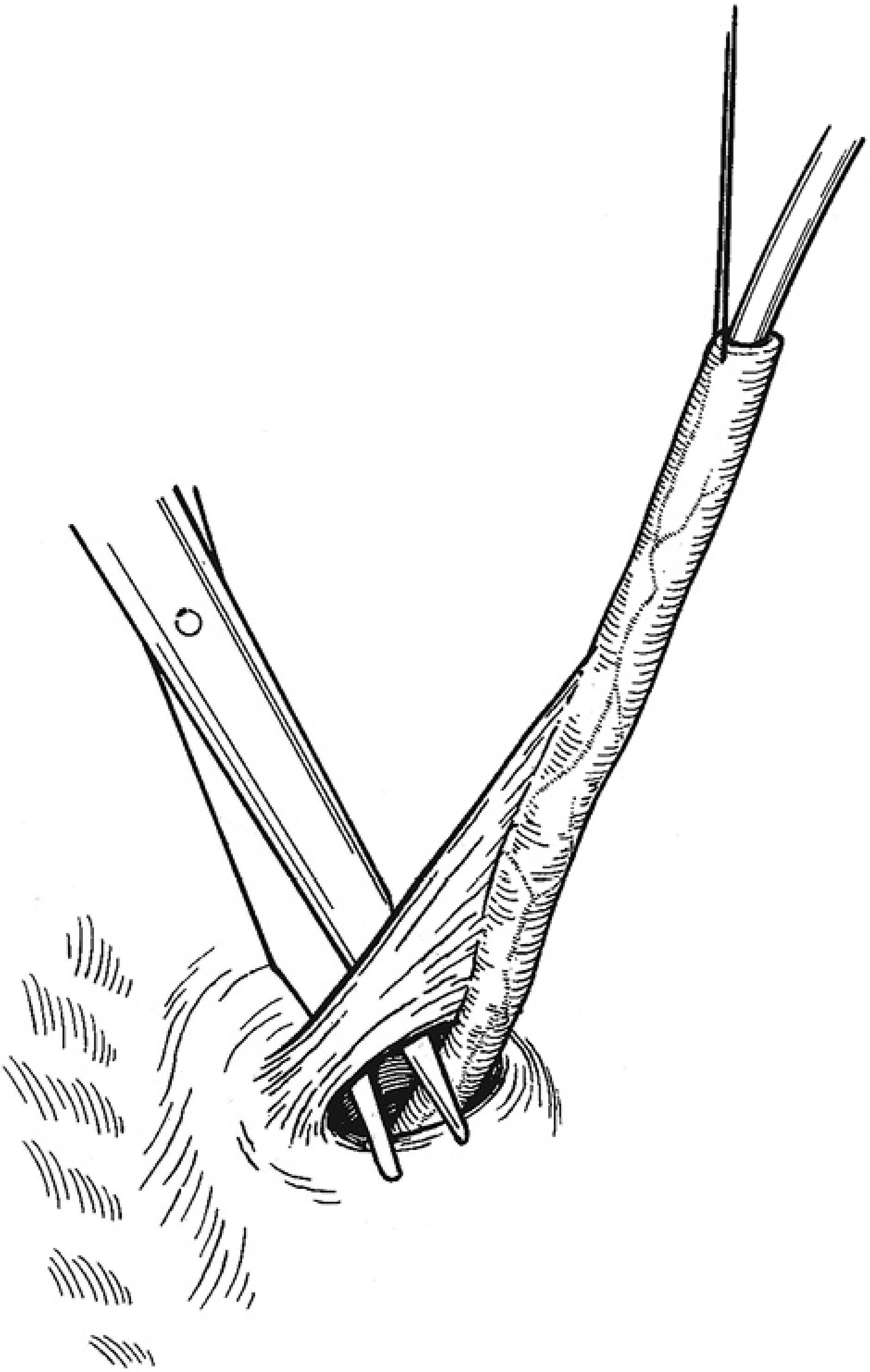
69.2a,b

The trigone is exposed and an infant feeding tube (3, 4, 6, or 8 Fr) is inserted into each ureter. A stay suture (5/0) is placed around each ureteric orifice. The assistant holds this stay suture with mild traction. The ureteric orifice is circumcised with diathermy (cutting and coagulation should be very low) and mobilization of the distal 2 cm of ureter can be performed with diathermy alone (these 2 cm will be excised later) (**Figure 69.3**).



69.3

It is essential to enter the correct plane between the bladder and the transparietal ureter, commencing below the orifice. Sharp scissors should be avoided, and Reynolds scissors make this procedure much easier. The tip of the scissors elevates the muscle fibers that attach the ureter to the bladder musculature (**Figure 69.4**). These fibers are grasped with fine forceps, coagulated, and divided. The dissection continues progressively, circumferentially until the ureter is completely free. Coagulation of the fibers should be carried out some distance from the ureter to avoid damaging its blood supply. The peritoneum is visible at the end of this dissection and should be teased away from the ureter. In boys, the vas deferens may lie close to the ureter at this point and care must be taken to avoid damaging it. A similar procedure can be used for the opposite ureter. In cases of ureteric duplication, both ureters are dissected together and should not be separated, thus avoiding damage to their blood supply.

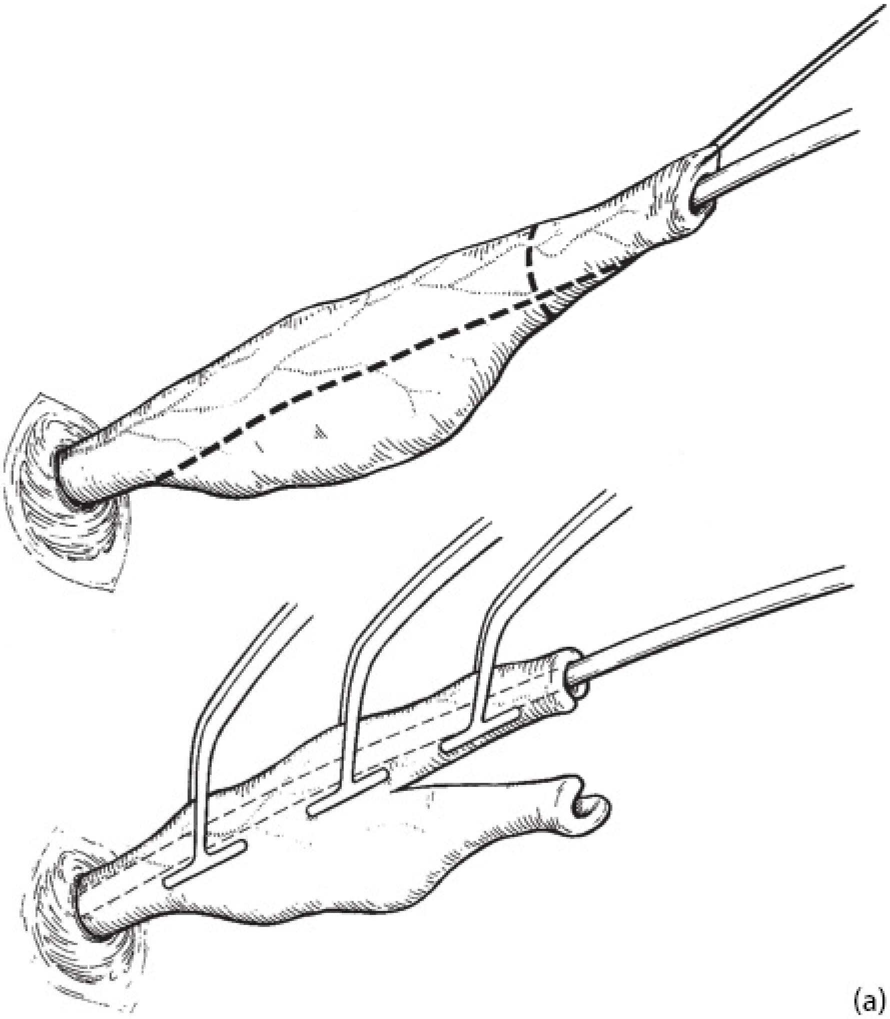


69.4

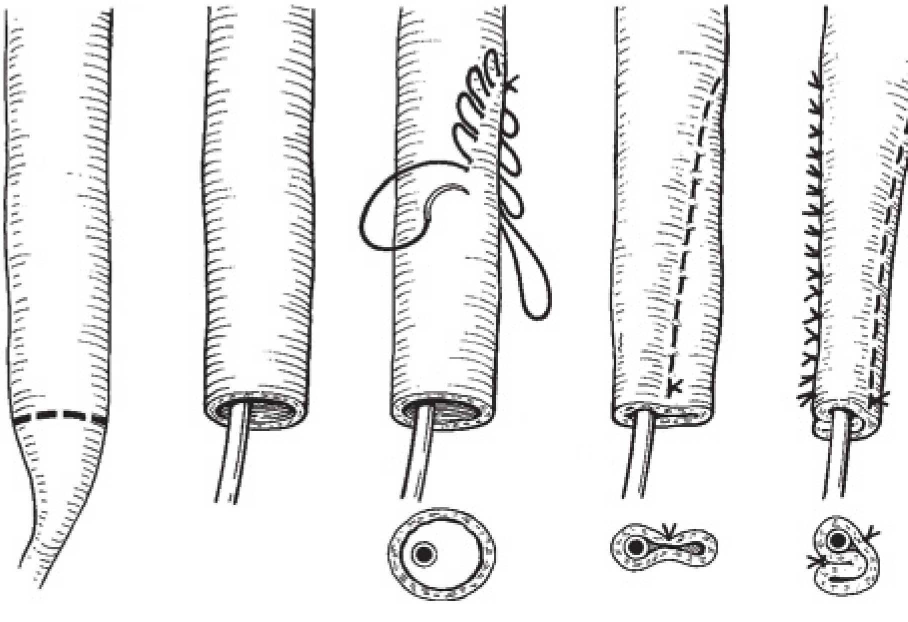
COHEN OR TRIGONAL REIMPLANTATION OF THE URETER

In some cases, the ureteric hiatus is wide and should be narrowed by one or two absorbable sutures. This is done to prevent the formation of a diverticulum. These sutures should narrow the hiatus, but still allow the free movement of the ureter and not restrict or constrict it.

The submucosal tunnel is then constructed. It is usually a horizontal tunnel, crossing the midline of the posterior surface of the bladder, just above the trigone. Its length should be at least four times the ureteric diameter (Paquin's rule) and, if this condition cannot be fulfilled, trimming or remodeling of the ureter should be considered (**Figure 69.5**). The site of the new ureteric orifice is selected and the bladder mucosa is lifted from the underlying bladder muscles with a pair of Reynolds scissors, starting either from the hiatus or from the new ureteric orifice (**Figure 69.6**). The tunnel should be wide enough to allow easy insertion of the ureter without constriction.

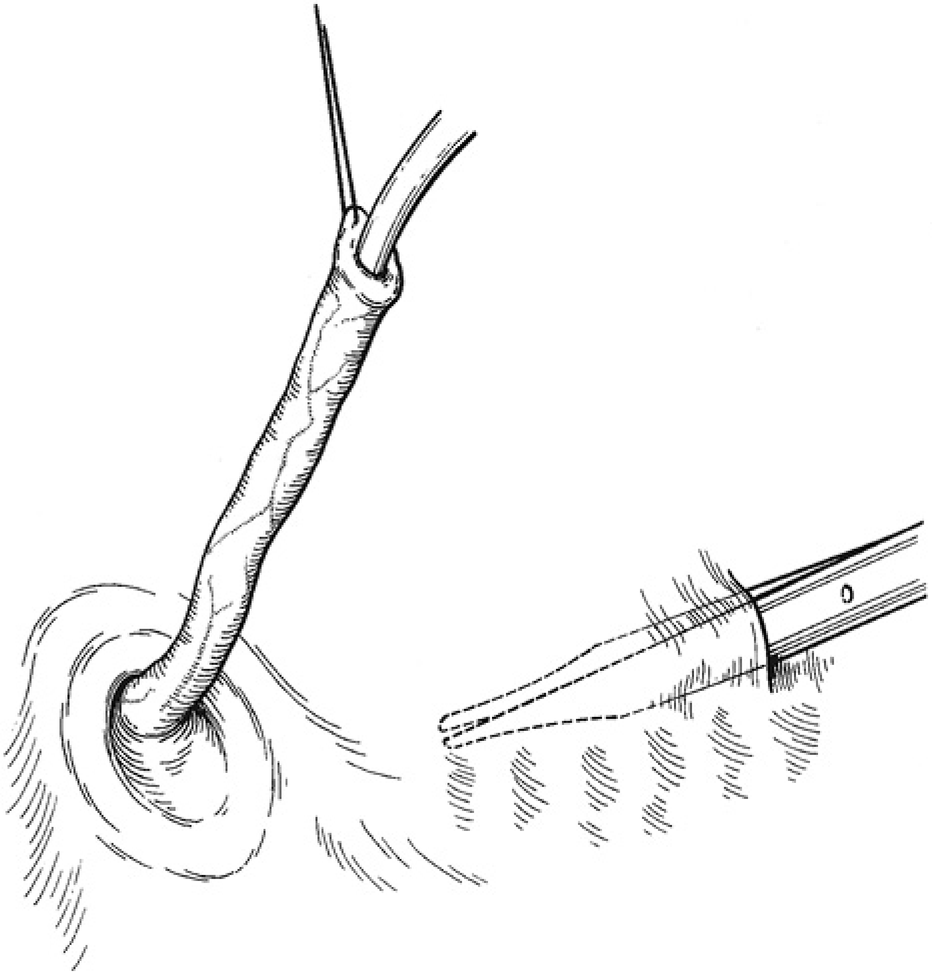


(a)



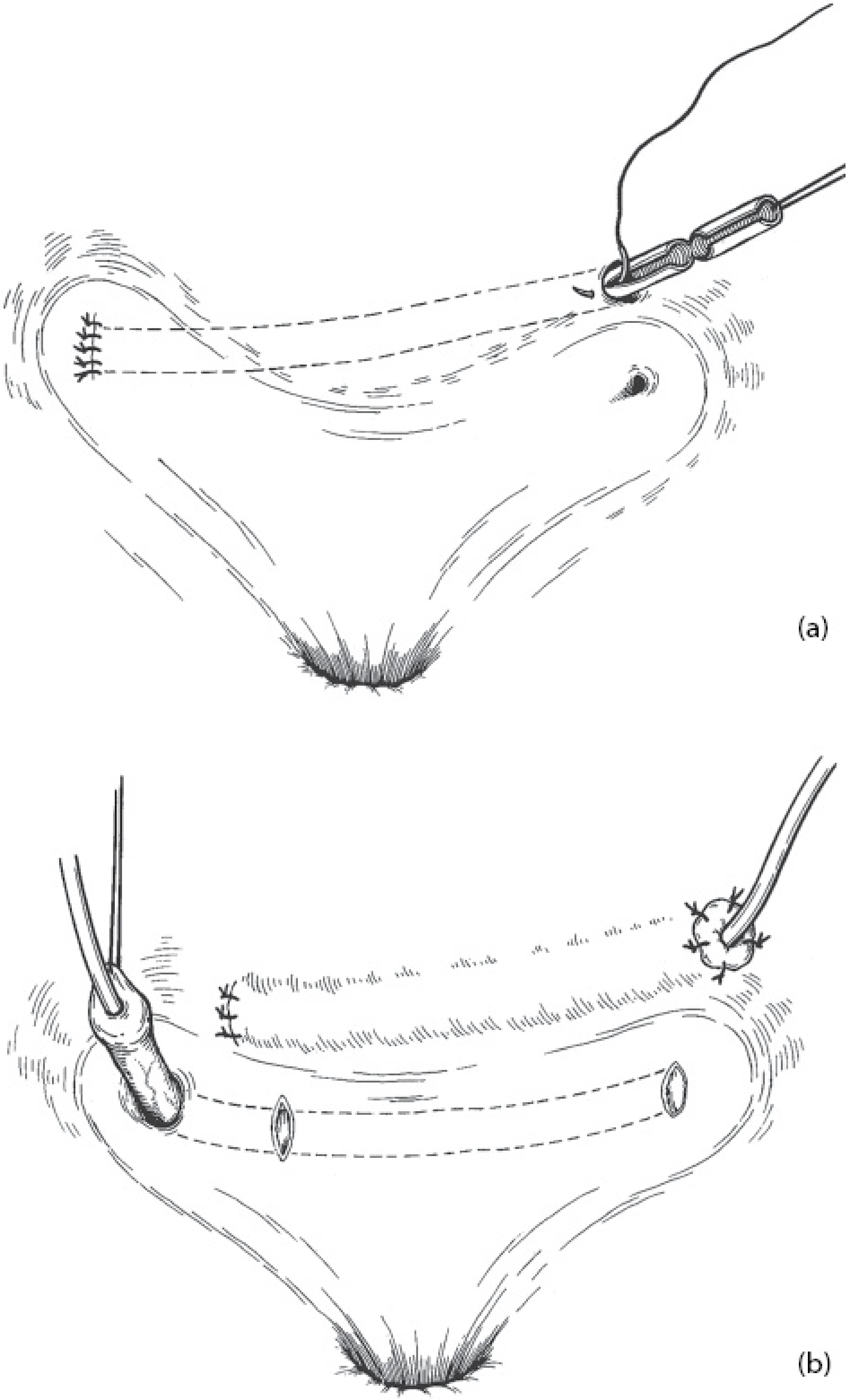
(b)

69.5a,b



69.6

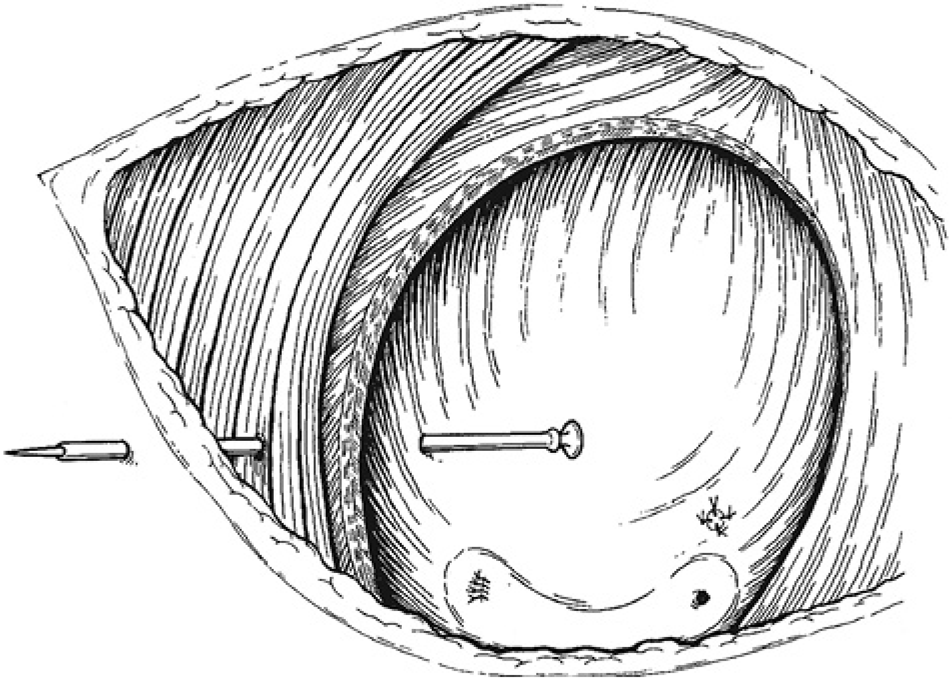
A similar procedure can be carried out for the opposite ureter in cases of bilateral reimplantation. The construction of the lowest tunnel that crosses the trigone can cause bleeding and the lifting of the mucosa is slightly less easy. A pair of artery forceps or rightangled forceps is inserted through the tunnel, and the stay suture is grasped and gently pulled to draw the ureter into place, taking care not to twist or kink it in the process (**Figure 69.7a**).



69.7a,b

The last 2 cm of ureter are excised and the ureteric opening is spatulated with a pair of angulated Potts scissors. The 5/0 absorbable suture anchors the ureter to the bladder muscles and the ureterovesicostomy is completed with interrupted 6/0 absorbable sutures (**Figure 69.7b**).

An infant feeding tube or a modified JJ stent (multipurpose Blue stent□; Bard Angiomed, Crawley, UK) is inserted into the reimplanted ureter and exteriorized through the bladder wall, the rectus muscle, and the skin (**Figure 69.8**). The feeding tube is left in position for 2 days, or for 10–15 days if the ureter has been remodeled. Alternatively, a retrievable JJ stent can be used. There is no consensus on the efficacy of drainage of the reimplanted ureter and some authors do not leave any drain.



69.8

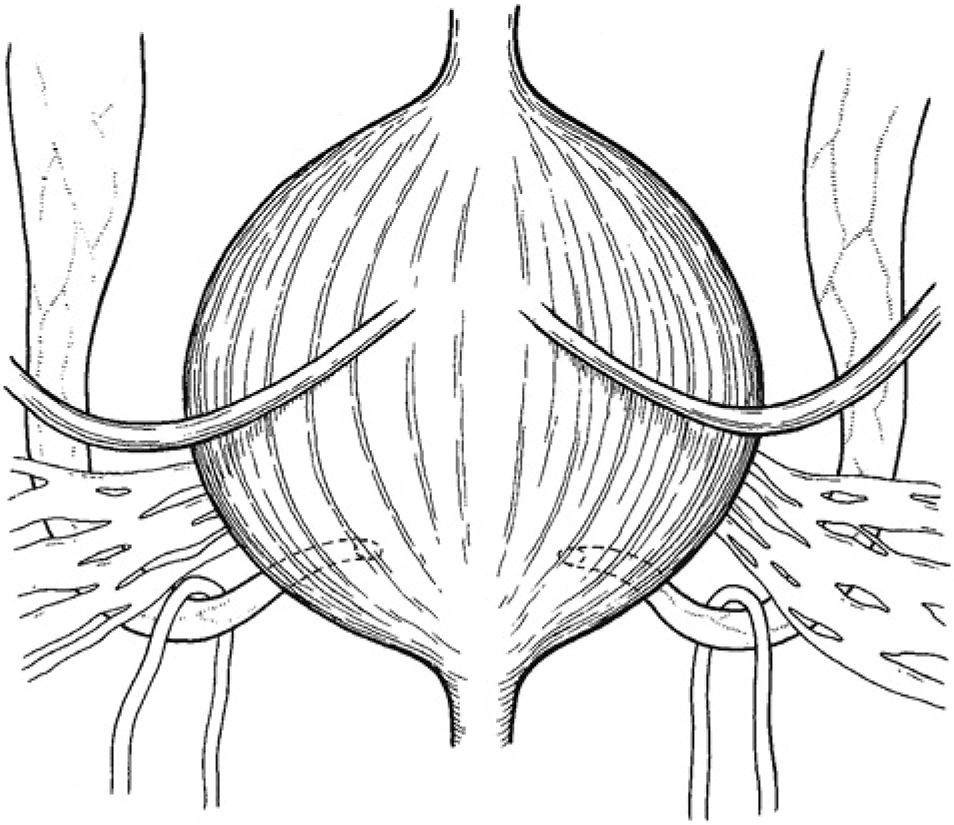
The bladder is drained either by a transurethral catheter, which is left *in situ* for 5 days, or by a suprapubic catheter. The bladder is closed with a 3/0 or 4/0 resorbable suture. The prevesical and subcutaneous spaces are drained by a suction drain. The abdominal wall, the subcutaneous tissues, and the skin are then closed.

Suprahiatal reimplantation of the ureter

Megaureters are the principal indication for this technique, which should be performed by an experienced pediatric urologist. It is a difficult procedure, which carries a significant complication rate.

The approach to the bladder, the retraction with the Denis Browne retractor, and the exposure of the bladder mucosa are as in the transhiatal procedure. The extravesical approach to the ureter is the main step in this procedure.

The peritoneum covering the dome and the lateral face of the bladder should be pushed upward, which involves ligation of the obliterated hypogastric ligament. It is then easy to mobilize the peritoneum upward and to expose the full length of the iliac vessels. The vas deferens and its pedicle are easily located and should also be freed before the ureteric reimplantation (**Figure 69.9**).



69.9

The ureter is now identified passing over the iliac vessels close to their division into the external iliac and hypogastric arteries. The ureter is progressively mobilized from this point down to the bladder, preserving its blood supply and the blood supply of the bladder. There is, in fact, a plethora of small vessels and nerves arising from the pelvic pedicles to the bladder, which cross the distal part of the extravesical ureter and should be preserved. In severe megaureters, the ureter is grossly dilated and kinked, and its dissection should be meticulous to straighten it and maintain enough tissue around it to preserve its blood supply and innervation. The ureter is divided at its entrance into the bladder and a stay suture facilitates its mobilization. The ureter, which normally passes under the vas deferens, should be redirected over it to straighten it out.

The distal segment of the ureter is usually narrowed and its excision allows urine to flow out freely. The ureteric diameter rapidly contracts and it is then possible to decide whether ureteric reimplantation can be achieved with or without remodeling or trimming. This decision is dictated by Paquin's rule: The length of the submucosal tunnel should represent at least four times the ureteric diameter.

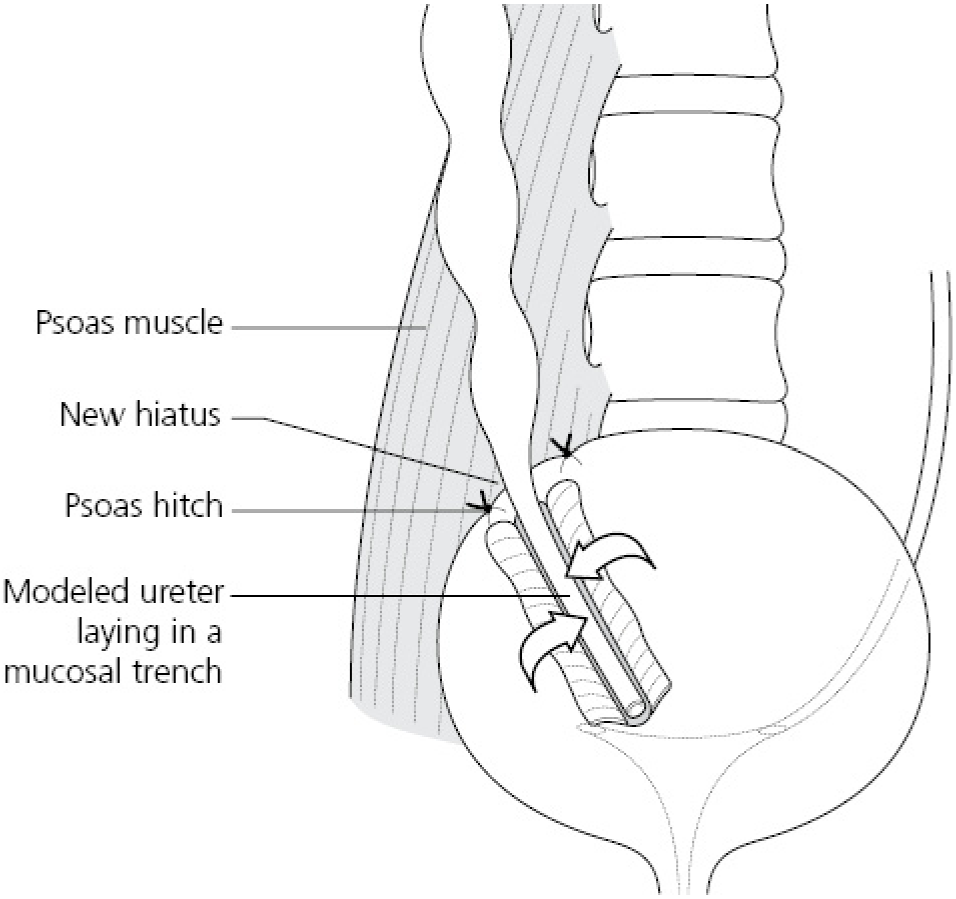
In the *Puigvert technique* (pull-through technique), the ureter is divided in a healthy zone and pulled into the bladder where it is attached by a direct circular anastomosis at the site of the original meatus. This classic direct ureteroneocystostomy is applied essentially to treat lesions of the pelvic ureter in its last 5 cm.

If the ureter remains too large after excision of its distal end, its caliber should be reduced, either by excising a strip of ureter (*Hendren's technique*) (see **Figure 69.5a**) or by infolding the ureter (*Kalicinski's technique*) (see **Figure 69.5b**).

Excision of a strip of ureter may threaten the ureteric blood supply, whereas ureter infolding can create a certain degree of obstruction. Whichever technique is chosen, the length of the remodeled or trimmed segment of ureter should not exceed the length of the submucosal tunnel.

REIMPLANTATION CREATING A NEW HIATUS

The entry hiatus of the ureter into the bladder should be medial and high at the top of the posterior surface of the bladder. The ureter should not be constricted at this level, and it is necessary to excise a disk of bladder to allow free passage of the ureter. The submucosal tunnel is fashioned, as described above, and should be vertical. Its distal end should open on the trigone. The passage of the freed ureter through the tunnel is the most difficult step of this procedure. The ureter should not be twisted or kinked, especially at the entrance into the bladder, and its pelvic course should be smooth. A few absorbable sutures are placed at its entrance into the bladder and sometimes the bladder itself is tacked down on the psoas muscle to maintain the smooth course of the ureter (**Figure 69.10**). The ureterovesicostomy is as described above. A bilateral procedure may be performed, although a bilateral extravesical approach of the ureters may affect bladder innervations and cause transient bladder dysfunction. Some authors prefer to perform a transureteroureterostomy to avoid bilateral suprahiatal reimplantation.



69.10 Gough's modification of the suprahiatal reimplantation of the ureter in case of megaureters.

Closure and drainage are as described above, except that the ureteric stent is maintained for at least 10 days if the ureter has been remodeled.

POSTOPERATIVE CARE

The child is usually hospitalized for 5 days. The ureteric stent is removed after 2 days (or 10 days if the ureter has been remodeled). The bladder catheter is removed on the fifth postoperative day. Both suction drains are usually removed on the second day. Bladder spasms are common, and administration of oxybutinin can be useful to reduce the discomfort. Antibiotic prophylaxis is a possible option with trimethoprim with or without sulfamethoxazole (co-trimoxazole).

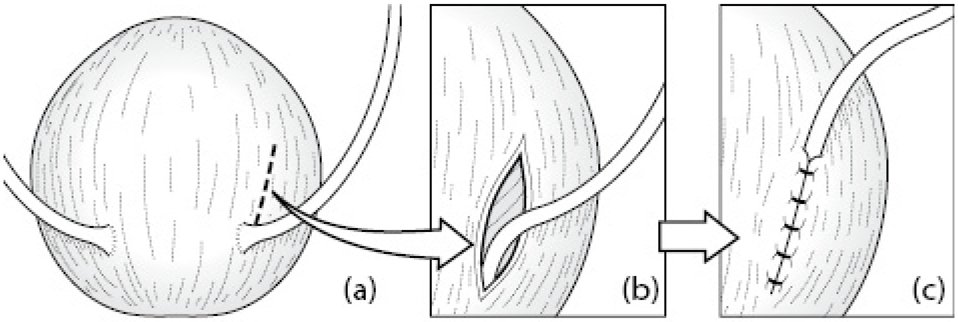
A US scan is recommended 3 months after surgery to ensure that there is no increased dilatation of the ureters. If the child is clinically well, control of absence of VUR is no longer necessary after surgery as a micturating cystogram is a rather unpleasant investigation with significant radiations and a risk of secondary UTI. If the kidney was damaged at the time of diagnosis, a repeated DMSA scan a few months after surgery might be useful.

COMPLICATIONS

Potential complications of open ureterovesico reimplantation are ureteral obstruction, febrile urinary infection and wound infection. After unilateral reimplantation, VUR can appear on the contralateral side. After ureteral modeling, ureteric stenosis and urine leakage are other potential complications requiring prolonged drainage and sometimes reintervention.

Extravesical reimplantation of the ureter: Lich-Gregoir technique

This technique consists of creating a submucosal tunnel by incising obliquely the detrusor muscle from the ureteric entry for 4–5 cm, laying the ureter inside the muscular trench, and suturing the muscular edges over it (**Figure 69.11a–c**). This technique, which can be performed either open or laparoscopically, implies a dissection of the posterolateral aspect of the bladder which, when performed bilaterally, may cause transient bladder dysfunction. The main advantage of this technique is that it avoids the opening of the bladder and reduces postoperative bladder spasms.



69.11a–c

OUTCOME

It is essential to reiterate that these procedures only aim at stopping VUR. All open approaches in experienced hands have a high success rate of more than 95%. The Cohen procedure permits the creation of a longer tunnel and reduces the risk of ureteral obstruction, but it makes further endoscopic procedures more difficult. The Lich- Gregoir technique is less invasive, but this is associated with a higher risk of urinary retention in bilateral repairs. The suprahiatal reimplantation is also associated with a risk of urinary retention caused by neurovascular injury during ureteral or bladder dissection.

MINIMALLY INVASIVE SURGERY

OPERATIONS

Lich-Gregoir technique: Laparoscopic approach

The patient is supine with the arms lying along the body. The surgeon stands at the head of the patient and the two assistants on each side. The screen and insufflator stand at the feet of the patient. A bladder catheter is placed and should be accessible during the procedure, and 3 mm instruments are used: Blunt grasper, bipolar forceps, hook, needle holder, scissors, and a 5 mm 30-degree optic.

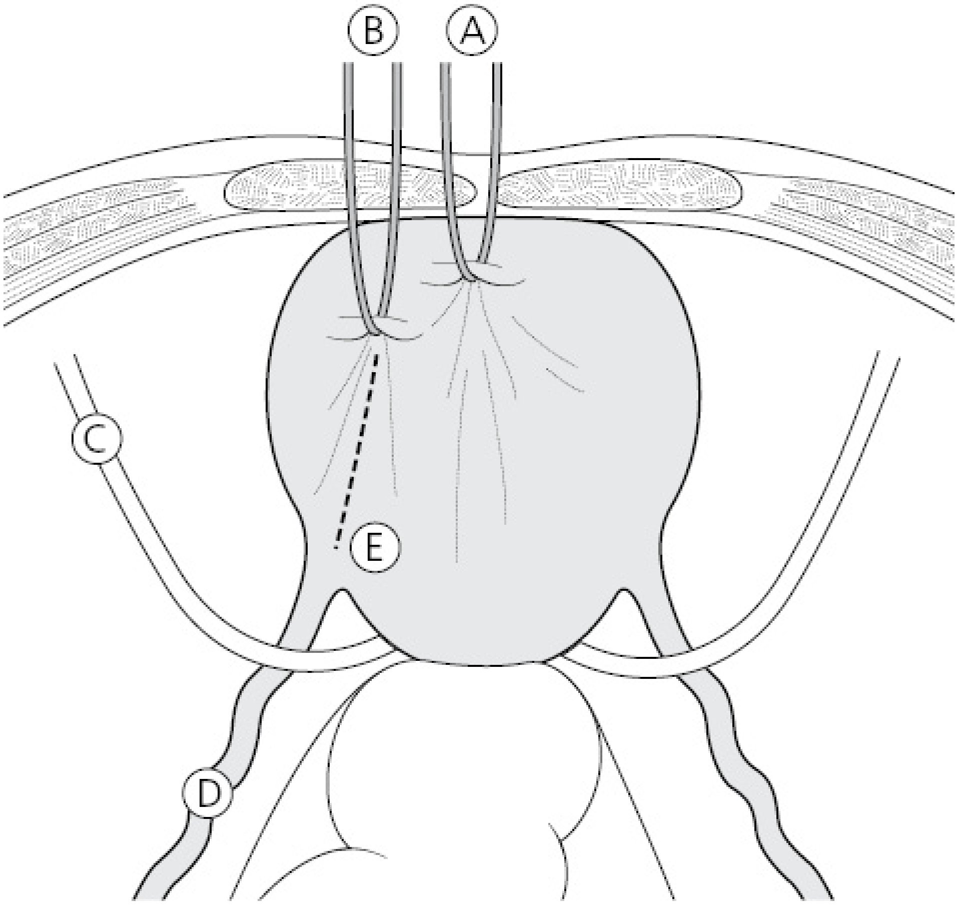
A transperitoneal approach is used. A 5 mm port is inserted with the open laparoscopy technique to avoid visceral damage. One 3 mm trocar is inserted in the left and one in the right flank, at the umbilicus level in children aged more than 2 years and lower in smaller children.

With the bladder empty, the ureter is easily identified where it crosses the external iliac vessels. The peritoneum is opened down to the VUJ. To avoid excessive handling of the ureter, a soft band is wrapped around the ureter and used for manipulation. In boys, the vas deferens is teased away from the ureter. In girls, the dissection is pursued through the broad ligament with preservation of the uterine artery. A 4–5 cm length mobilization of the ureter is needed.

The bladder dome is suspended to the anterior abdominal wall with a transcutaneous stay suture in order to expose the VUJ.

DETRUSOROTOMY

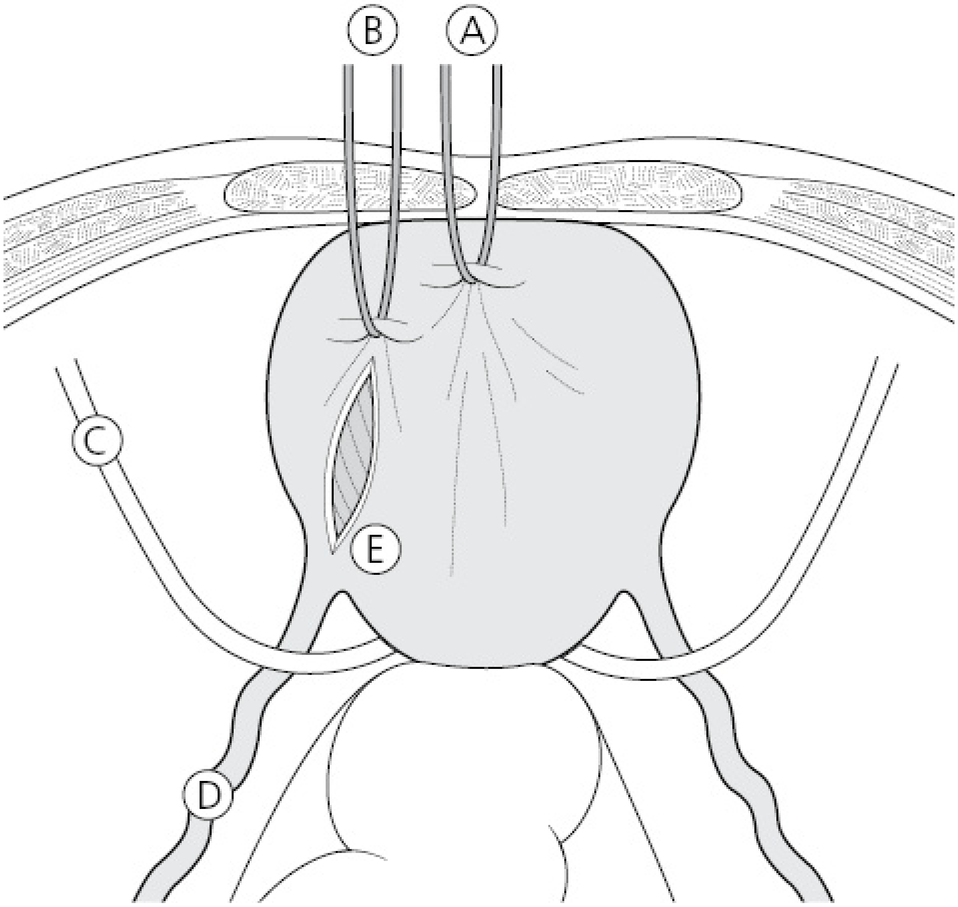
The direction and length of the muscular incision are outlined with the unipolar coagulation following Paquin's rule (**Figure 69.12**: A, bladder suspension; B, suspension of the distal end of the detrusorotomy; C, vas deferens; D, ureter; E, incision line of the detrusor).



69.12

A second stay suture is placed at the tip of the incision. The bladder is filled to create a bulging out of the mucosa. The muscular fibers are divided with the scissors (see **Figure 69.16**). Lateral dissection of the ureter and bladder should be limited to avoid damage to pelvic nerves. We recommend a gentle soft tissues dissection around the lower ureteral part and no extensive coagulation.

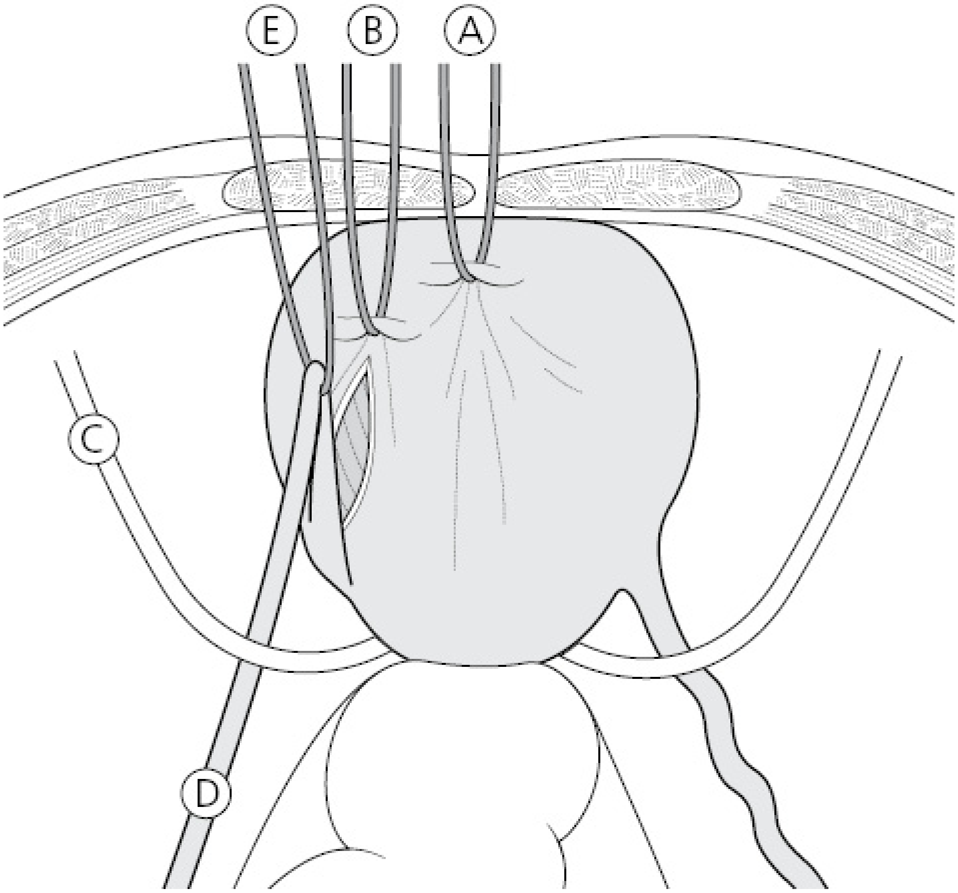
In case of mucosal tear, we favor closure by an ENDOLOOP®, rather than direct suture (**Figure 69.13**: A, bladder suspension; B, suspension of the distal end of the detrusorotomy; C, vas deferens; D, ureter; E, detrusorotomy).



69.13

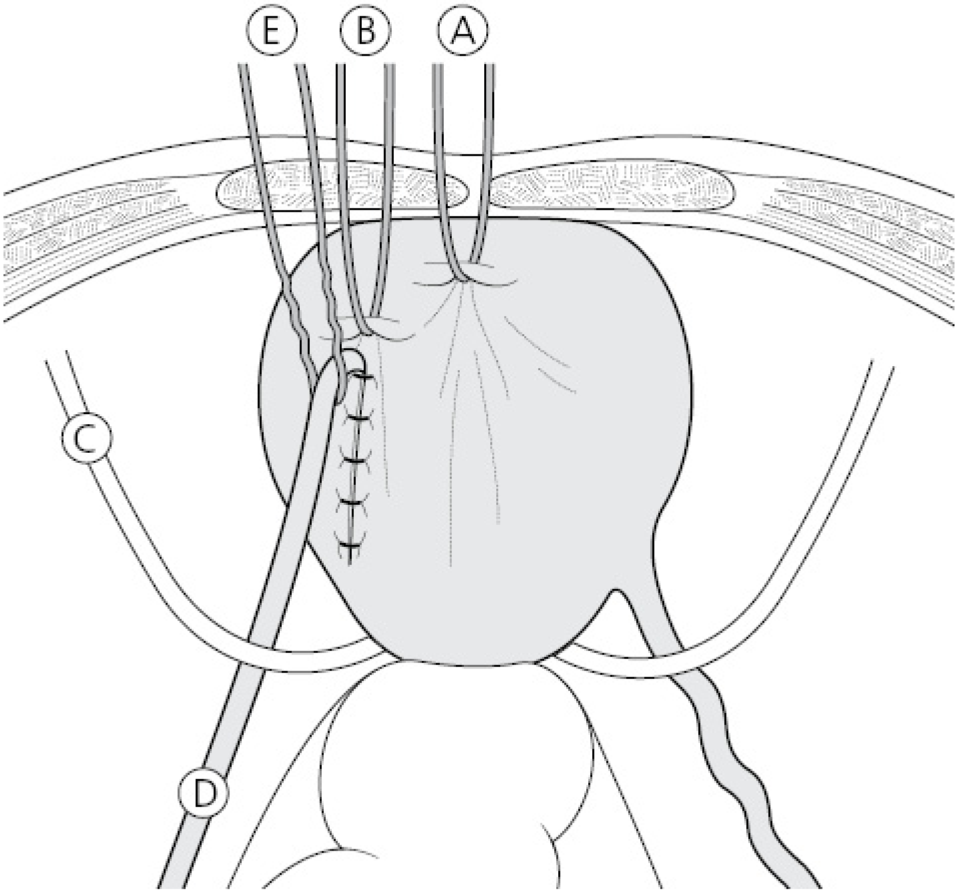
DETRUSORORRHAPHY

The ureter is laid between the two edges of the muscular trench and kept in this position with a third stay suture through the soft band used for mobilization of the ureter (**Figure 69.14**: A, bladder suspension; B, suspension of the distal end of the detrusorotomy; C, vas deferens; D, ureter; E, ureteric suspension).



69.14

The detrusor is then sutured over the ureter with three or four stitches of 3/0 or 4/0 non-resorbable suture. Particular attention should be given to avoid narrowing the entry of the ureter into the trench (**Figure 69.15**: A, bladder suspension; B, suspension of the distal end of the detrusorotomy; C, vas deferens; D, ureter; E, ureteric suspension).



69.15

BILATERAL REIMPLANTATION

The same procedure can be done on both sides with the same approach. In case of ureteral duplication, both ureters are dissected and laid into the trench together.

POSTOPERATIVE CARE

Unilateral reimplantation following this technique can be performed as a day case, the patient being discharged after complete micturition. Only standard painkillers are needed. For bilateral reimplantations, we prefer to keep the patient hospitalized until the next day, as the risk of urinary retention is higher. A US scan is performed at 1 month following the operation. As for open surgery, micturating cystogram is not routinely performed.

This technique significantly reduces postoperative bladder spasms and adverse effects of bladder opening. Its success rate is comparable to all extravesical ureteric reimplantation. The risk of transient bladder dysfunction following bilateral laparoscopic ureteral reimplantation seems to be less than after open procedures.

COMPLICATIONS

The potential complications of laparoscopic reimplantation are ureteral kinking or obstruction and febrile urinary infection. Urinary retention is more frequent after bilateral reimplantation and may require the insertion of a supra-pubic catheter. Another potential complication is ureteral injury or obstruction (ischemia) owing to excessive handling of the ureter or an excessive closure of the new channel.

Laparoscopic reimplantation of a dilated megaureter

Although technically challenging, the reimplantation of a dilated megaureter is also possible using the laparoscopic approach. The initial steps of the procedure are the same as previously described. Once the distal part of the ureter is completely dissected, the bladder is filled with air or saline solution. The detrusor muscle is cautiously divided down with scissors until the mucosa is exposed for 4–6 cm. The distal ureter is ligated with a 2/0 resorbable suture and the ureter divided proximal to it. The distal end of the ureter is excised and sent for histological assessment. Subsequently, the free ureteral end is delivered out through the ipsilateral 3 mm port. The lower end is tailored over a 10 Fr tube. A JJ stent is placed. Later the entire assembly is carefully replaced in the abdomen. The pouting mucosa at the distal-most part of the incision is opened and mucosa-to-mucosa ureterovesico anastomosis is completed with 6/0 resorbable sutures. The ureter is placed in the newly created tunnel and the detrusor muscle is reapproximated with three or four separate intracorporeal stitches with non-absorbable sutures 3/0. At the final stage, the ureter is mobilized without tension to avoid ectasia or ischemia and the urine catheter is removed. The JJ stent is removed a few weeks later by endoscopy.

Extravesical robotic-assisted laparoscopic ureteral reimplantation

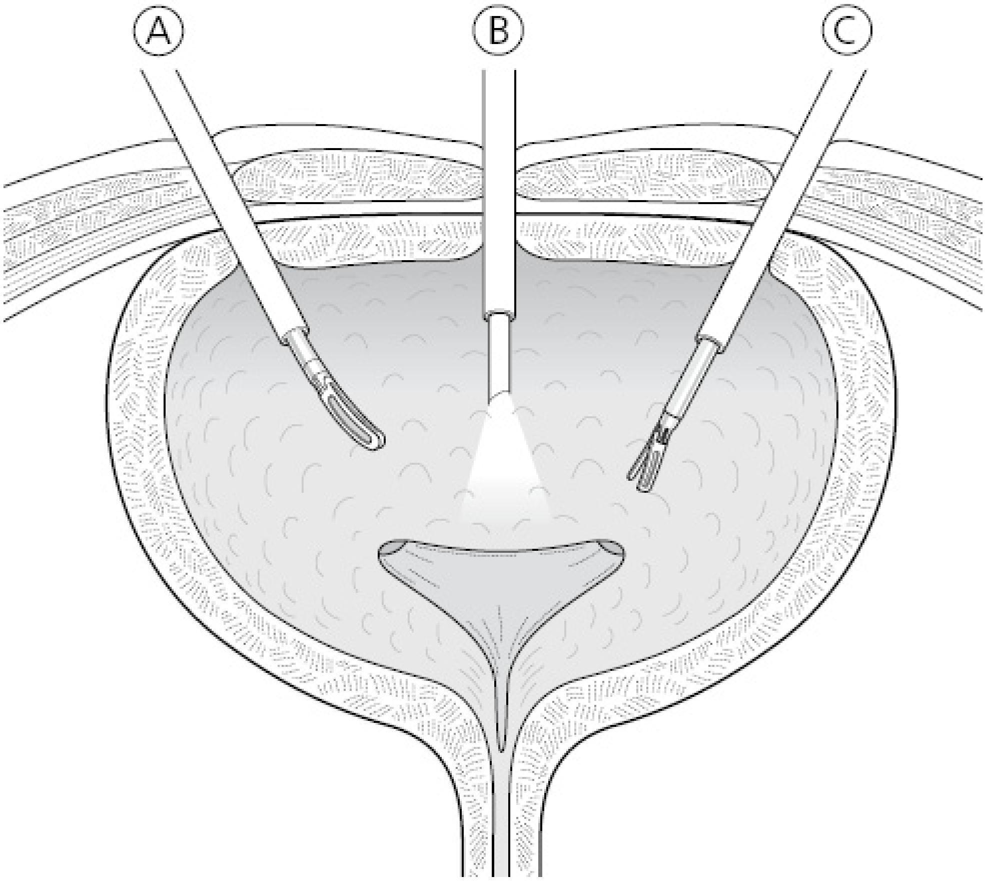
Extravesical robotic-assisted laparoscopic ureteral reimplantation is an alternative to open and laparoscopic surgery. Trocars sizes are 12 mm or 8.5 mm for camera and 8 mm or 5 mm for instruments. Robotic surgery has many advantages, notably viewing quality and high precision of motion. However, larger trocars size can be a problem even if incidence of port-site complications remains low. Major disadvantages of robot-assisted surgery are significantly longer operating time and higher costs.

Although robotic-assisted treatment of VUR is a promising technique, its advantages and disadvantages over open or laparoscopic techniques have yet to be evaluated.

Vesicoscopic Cohen reimplantation of the ureters

For this procedure the patient is in a cystoscopy position with access to the urethral meatus. In children under 5 years of age, the position of the surgeon is the same as described for the laparoscopic Lich-Gregoir technique.

A cystoscopy is performed to monitor the placement of the 5 mm trocar inserted through a midline suprapubic incision. The bladder is filled and the dome is suspended to the anterior abdominal wall as described before. Two 3 mm or 5 mm trocars are inserted through each side of the bladder with a concomitant endoscopic control. The cystoscope is then removed and the CO_2 insufflation is started with the median trocar with a pressure of 8–12 mmHg (**Figure 69.16**: A, C, operating ports; B, optical port).



69.16

TRANSHIATAL URETERAL DISSECTION

A 4 cm ureteral stent is inserted into each ureter and stitched to the ureteral meatus with a 5/0 resorbable suture. The ureteral dissection follows the Cohen technique using the hook and a pledget. The ureteral hiatus is narrowed with several stitches to avoid CO_2 leakage outside the bladder. The ureter is stitched to the detrusor muscle to avoid its retraction, as with the open procedure.

URETERAL REIMPLANTATION

Subcutaneous tunnels are fashioned with the scissors and each ureter is pulled under the bladder mucosa using a stay suture and/or a ureteral stent. The extremity of each ureter is excised, spatulated, and stitched to the detrusor with 5/0 and 6/0 resorbable sutures. The mucosa is stitched as with the open procedure.

The trocar orifices are closed under endoscopic control. The gas is removed via a transurethral catheter and left in place for 48 hours.

POSTOPERATIVE CARE

The child can go home 48 hours after surgery with the same advice as that given before.

The success rate is equivalent to the open Cohen procedure with much less postoperative discomfort. However, this technique is more challenging and the learning curve is longer.

ENDOSCOPIC TREATMENT

HISTORY

Endoscopic treatment of a bulking agent was pioneered by Matouschek in 1981 and further developed and popularized by O'Donnell and Puri in 1984. Since the American Food and Drug Administration (FDA) approval in 2001 of dextranomer/hyaluronic acid copolymer (Deflux® [Palette Life Sciences, Stockholm, Sweden]) as a tissue-augmenting substance for subureteral injection, endoscopic treatment has become a widely accepted minimally invasive alternative to long-term prophylaxis and open or laparoscopic surgical treatment in the management of VUR.

PRINCIPLES AND JUSTIFICATION

The principle of the endoscopic treatment of VUR is to coapt the ureteral orifice and detrusor tunnel by submucosal injection of a bulking agent. This provides a solid support under the refluxing ureteral orifice, which improves the valve mechanism stopping urine from refluxing into the ureter. This technique is minimally invasive and technically straightforward, with a relatively short learning curve and low complication rate.

BULKING AGENTS

Various injectable agents can be used for this purpose. The bulking agent is degraded in time and replaced by an autologous collagenic matrix, which induces a controlled iatrogenic scar responsible for the therapeutic effect. Polytetrafluoroethylene (PTFE), also called Teflon®, was the first material investigated for endoscopic treatment of VUR. Despite good long-term outcomes, PTFE has been nearly abandoned as a result of information regarding the risk of particle migration and the success of other bulking agents.

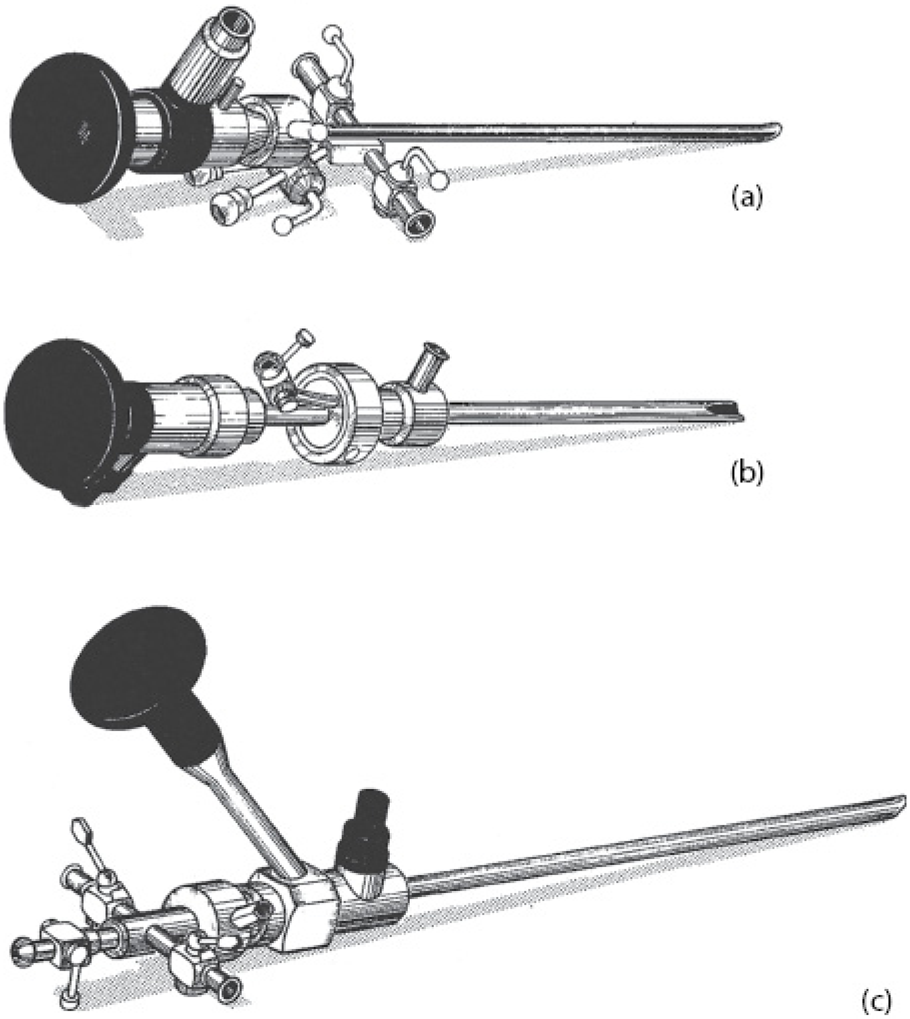
Over the past two decades, the use of dextranomer/hyaluronic acid (Dx/HA) microspheres (Deflux®; Dexell®– Istem Medikal, Ankara, Turkey) has been widely used with good success rates. Poly-dimethyl-siloxane (Macroplastique®; Laborie Medical, Williston, VT, USA) is an alternative to Dx/HA with comparable success rates.

Polyacrylate polyalcohol copolymer (Vantris® [Promedon, Cordoba, Argentina]), a non-biodegradable substance of synthetic origin belonging to the acrylic family, has also been available since 2010.

INJECTION TECHNIQUE

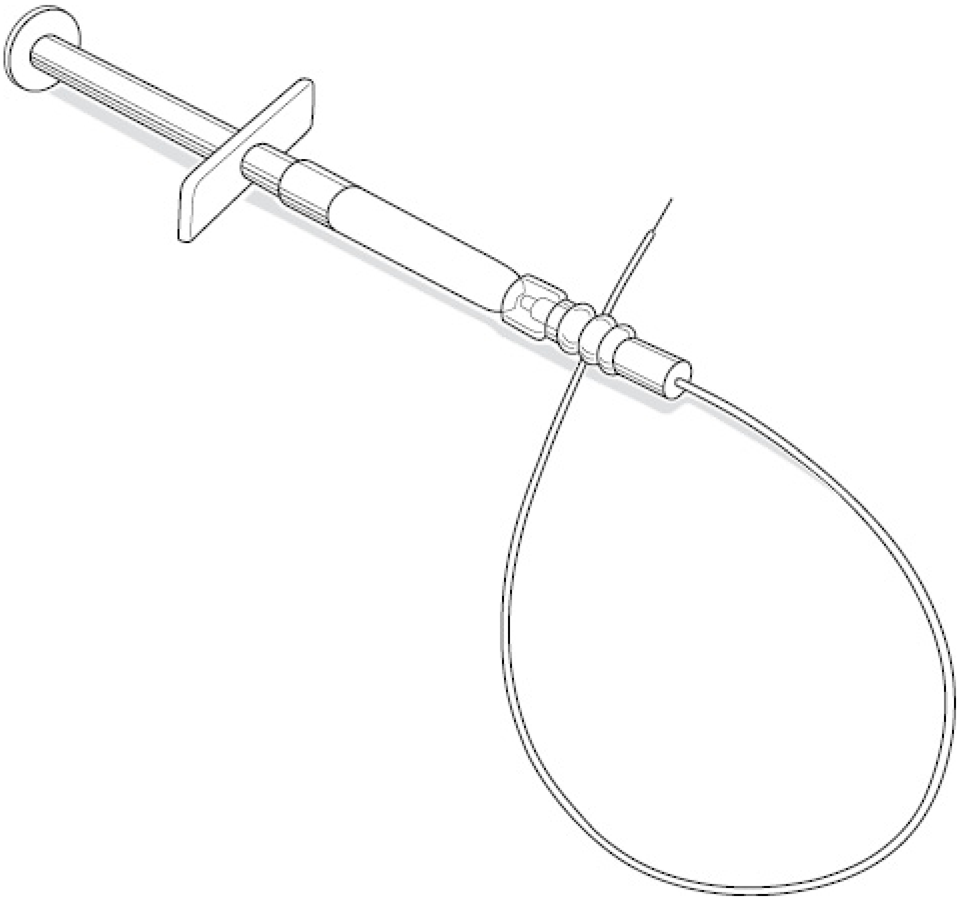
The child is placed in a lithotomy position under general anesthesia.

Cystoscopy is performed with a cystoscope of 9.5 Fr size or higher (**Figure 69.17a–c**). All cystoscopes available for infants and children can be used for this procedure, but an angled cystoscope permits direct passage of the needle in line with the ureter without bending the needle.



69.17a–c

Several needles are available for injection. The disposable Puri catheter (Storz) is a 4 Fr nylon catheter onto which is swaged a 21 gauge needle with 1 cm of the needle protruding from the catheter. Alternatively, a rigid needle can be used. A 1 mL syringe prefilled with the bulking agent is attached to the injection catheter (**Figure 69.18**).



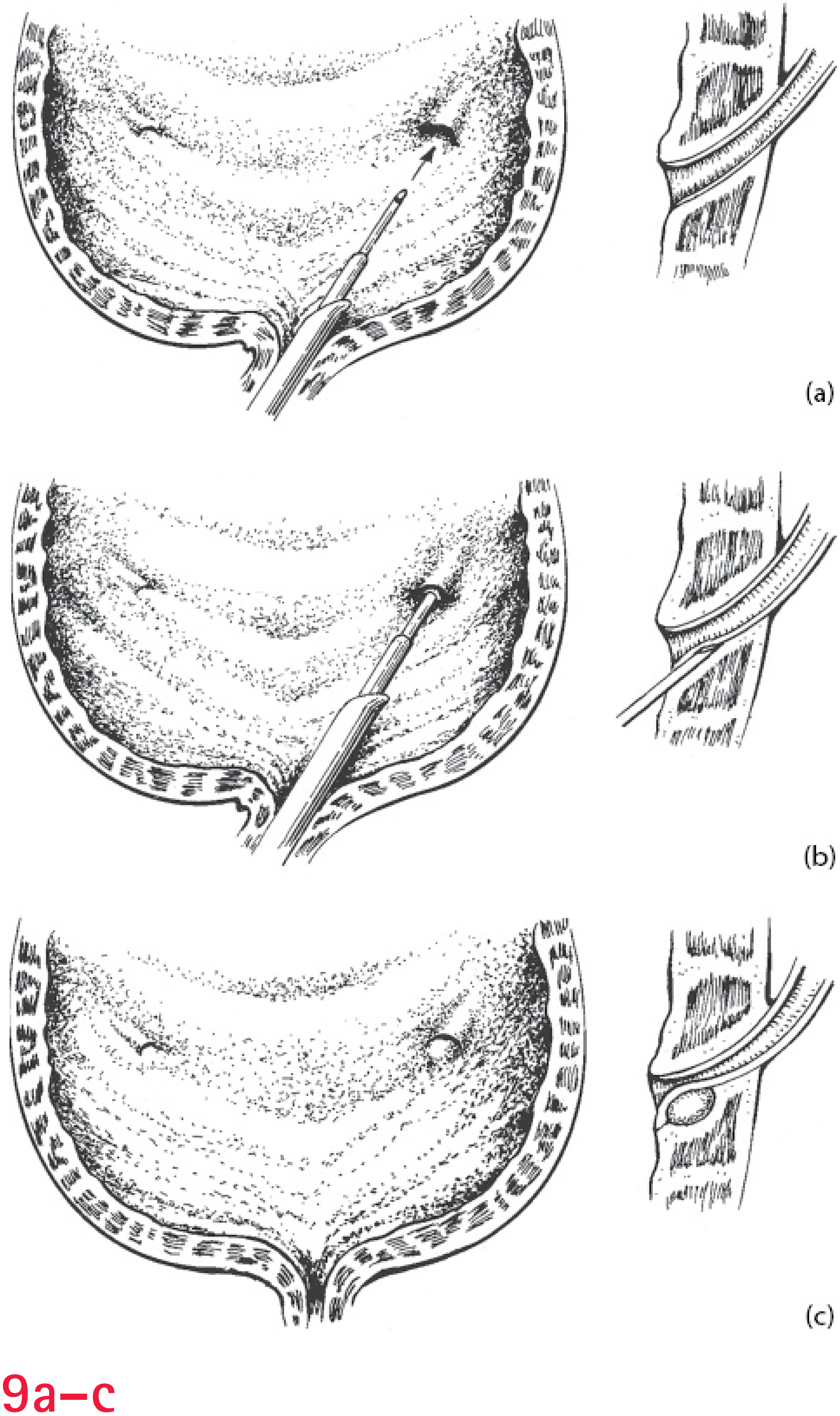
69.18

The bladder wall, trigone, bladder neck, and both ureteric orifices are inspected. The bladder should be almost empty before proceeding with injection, since this helps to keep the ureteric orifice flat rather than away in a lateral part of the field.

STING procedure: Subureteric Teflon® injection

In the traditional STING procedure, the needle is introduced under the bladder mucosa 2–3 mm below the refluxing orifice at a 6 o’clock position (**Figure 69.19a**). The needle is advanced about 4–5 mm into the lamina propria in the submucosal portion of the ureters and the injection started slowly (**Figure 69.19b**). The injection is continued until there is a prominent bulge and the ureteral orifice has assumed a crescent-like shape

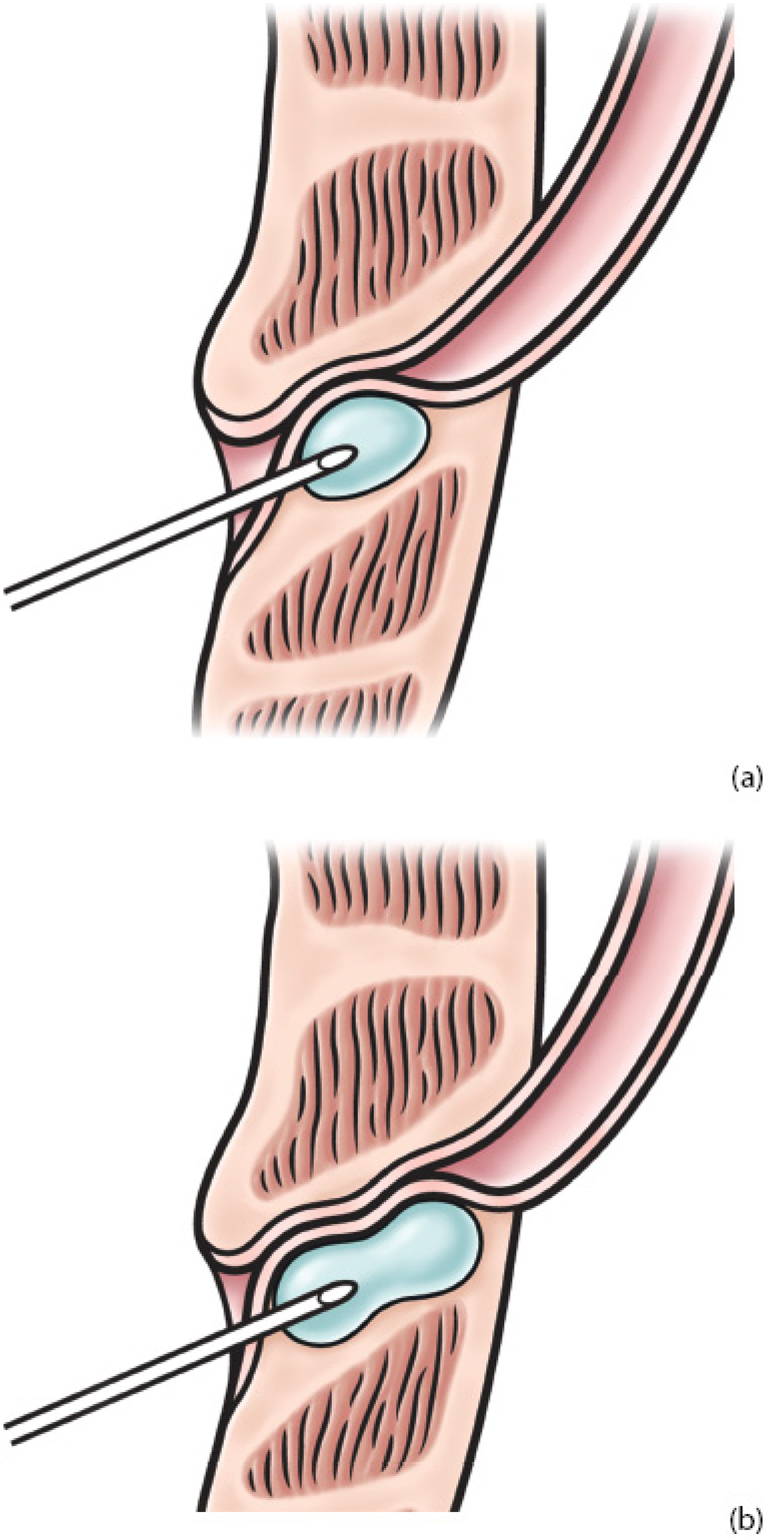
(**Figure 69.19c**). Most refluxing ureters require 0.4–1.0 mL of bulking agent to correct reflux.



69.19a–c

Hydrodistension implantation technique

The hydrodistension implantation technique (HIT) has been developed by Kirsch et al. (2004). Hydrodistention is performed with the tip of the cystoscope placed at the ureteral orifice. A pressured stream is achieved by placing the irrigation bag approximately 1 meter above the bladder. The needle is advanced into the ureteral tunnel and the bulking agent is injected along the entire length of the detrusor tunnel for maximal coaptation. Hydrodistension allows visualization of the intraureteral injection site and assessment of treatment progress (**Figure 69.20a**). The double HIT technique is a modification with two intraureteral injections that coapt both the ureteral tunnel and orifice, creating a "mountain range appearance" (**Figure 69.19b**).



69.20a,b

POSTOPERATIVE CARE

Postoperative urethral catheterization after an endoscopic procedure is not necessary. The majority of patients are treated as day cases. Co-trimoxazole is sometimes prescribed in prophylactic doses for 6–12 weeks after the procedure. A follow-up renal and bladder ultrasonographic scan is obtained a few months after endoscopic correction of reflux.

COMPLICATIONS

Complications after endoscopic treatment of VUR are rare. The significant complications with this procedure are treatment failure, recurrence of VUR, and rare cases of ureteral obstruction and material migration.

VIDEO

OPS_8e_video_69.1 Lich Gregoir reimplantation

FURTHER READING

Demède D, Cheikhelard A, Hoch M, Mouriquand P. Evidence-based médicine and vesico-ureteral reflux. *Ann Urol* 2006; 40: 161–74.

Kirsch AJ, Perez-Brayfield M, Smith EA, Scherz HC. The modified STING procedure to correct vesicoureteral reflux: Improved results with submucosal implantation within the intramural ureter. *J Urol* 2004; 171: 2413–16.

Puri P, Kutasy B, Colhoun E, Hunziker M. Single centre experience with endoscopic subureteral dextranomer/hyaluronic acid injection as first line of treatment in 1551 children with high-grade vesicoureteral reflux. *J Urol* 2012; 188: 1485–9.

Puri P, Pirker M, Mohanan N, et al. Subureteral dextranomer/hyaluronic acid injection as first line treatment in the management of high-grade vesicoureteral reflux. *J Urol* 2006; 176: 1856–60.

Soulier V, Scalabre A, Lopez M, et al. Laparoscopic vesico-ureteral reimplantation with Lich-Gregoir approach in children: Medium term results of 159 renal units in 117 children. *World J Urol* 2017; 35: 1791–8.

Valla JS, Steyaert H, Griffin SJ, et al. Transvesicoscopic Cohen ureteric reimplantation for vesicoureteral reflux in children: A single-centre 5-year experience. *J Pediatr Urol* 2009; 5: 466–71.

Yeung CK, Sihoe JD, Borzi PA. Endoscopic cross-trigonal ureteral reimplantation under carbon bladder insufflations: A novel technique. *J Endourol* 2005; 19: 295–9.

70

Ureteric duplication

SARAH L. HECHT AND DUNCAN T. WILCOX

INTRODUCTION

Ureteral duplication is common and leads to a variety of anatomic and functional anomalies. The anatomic sequelae of ureteral duplication include partial duplication, ectopic ureter, ureterocele, and vesicoureteric reflux. Clinical manifestations include urinary tract obstruction, infection, incontinence, and renal dysfunction. Severity varies widely, and treatment ranges from observation to major surgical reconstruction. Treatment, when indicated, is primarily surgical and is aimed at preservation of renal function, minimizing morbidity from urinary tract infection (UTI), correction of urinary incontinence, and relief of symptoms associated with urinary obstruction.

PRINCIPLES AND JUSTIFICATION

Renal duplication anomalies are the most common congenital urinary tract anomaly with an incidence of 0.8% noted on autopsy studies. A *duplex kidney* refers to a kidney with two moieties, upper and lower, each with an independent pyelocaliceal drainage system and may be partial or complete. The two collecting systems in partial duplication may join at any point before entering the bladder. Complete duplication occurs when double ureters drain their respective poles and empty separately into the genitourinary tract.

Duplication anomalies can present in one of several ways but nowadays most are diagnosed antenatally on the maternal ultrasound (US). Postnatally, they may be diagnosed as workup for UTI and about 10% who present with a UTI are found to have this. They may also present with urinary incontinence in toilet-trained girls. Children may also present with symptoms of urinary tract obstruction such as intermittent pain or failure to thrive, and hydronephrosis is typically noted on US.

There is no side predilection and bilateral duplication occurs in 20–40% of cases. Duplication is more common in females and Caucasians. It can be inherited as an autosomal dominant trait with incomplete penetrance; the incidence of duplication among members of affected families is 8%.

Embryology

Two developmental structures give rise to the mature human kidney and ureters; the ureteric bud, which develops into the collecting system and ureters, and the metanephros, which develops into the nephrons and renal parenchyma. The ureteric bud begins as an epithelial outpouching off the distal mesonephric (Wolffian) duct. Typically, there is a single ureteric bud on each side giving rise to bilateral single ureters. The ureteric bud grows into the metanephros and a process of reciprocal induction causes nephrogenesis and ureteral branching. The renal unit with its branched collecting system migrates cranially, and the ureter grows caudally to merge with the urogenital sinus (the eventual bladder). Once incorporated into the urogenital sinus, the ureteral orifice migrates in a cranial and lateral direction from the bladder neck to its orthotopic position on the trigonal ridge.

Embryopathology

Partial ureteral duplication occurs when a single ureteral bud branches prior to reaching the metanephros, whereas complete ureteral duplication occurs when two ureteral buds branch off the mesonephric duct. Each bud or branch induces maturation of an overlying renal unit. Distally, the lower ureter will be incorporated into the urogenital sinus first, and will migrate further craniolaterally. The upper ureter is incorporated later or not at all and is thus medially and caudally displaced in the bladder or, if it never reaches the urogenital sinus, is ectopically located along the mesonephric duct derivatives. The craniolateral ureteral orifice is thus reliably associated with the lower renal unit and the caudomedial ureteral orifice with the upper renal unit, an embryologic relationship known as the Weigert–Meyer law.

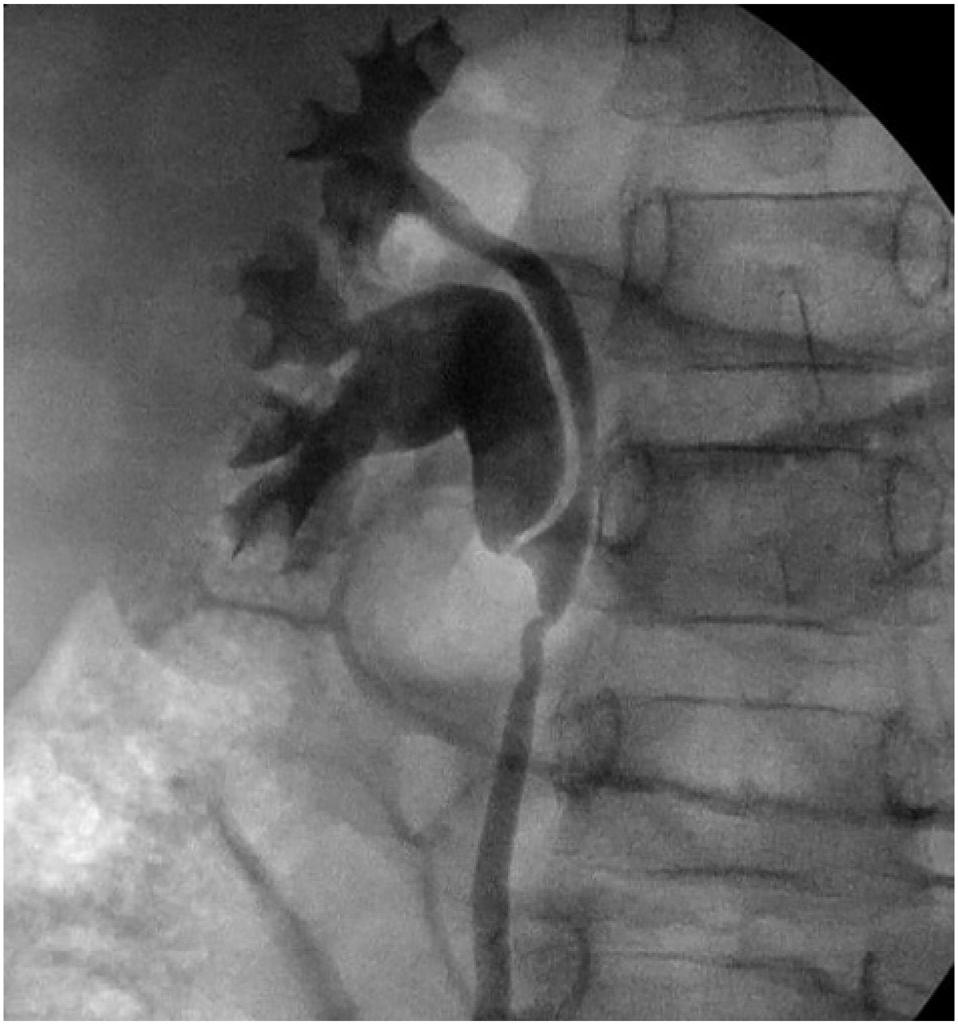
Table 70.1 lists ureteral duplication anomalies and their associated sequelae. Note that all of these anomalies can predispose to UTI.

Table 70.1 **Duplication anomalies and their associated sequelae**

Anomaly	Potential problems
Incomplete duplication	Lower-pole UPJ obstruction Ureteroureteral reflux
Ectopic ureter	Upper-pole obstruction Urinary incontinence Epididymitis
Ureterocele	Upper-pole obstruction Lower-pole obstruction Bladder outlet obstruction Urinary incontinence due to sphincter disruption
Vesicoureteric reflux	Reflux nephropathy

Incomplete duplication

Most ureteral duplications are incomplete and clinically insignificant. Partial duplication ranges from a bifid renal pelvis to a Y-type duplication with the ureters joining at any point prior to their insertion into the bladder (**Figure 70.1**). About 10% of the population has a bifid renal pelvis, and this should be considered a normal variant. One problem encountered with incomplete duplication is obstruction of the ureteropelvic junction (UPJ). This occurs almost exclusively at the lower pole, though several case reports of upper-pole UPJ obstruction exist. Lower-pole UPJ obstruction can occur with complete duplication but is reported more commonly with partial duplications. The intrarenal architecture in duplicated systems may explain the predilection of UPJ obstructions for the lower pole. Typically, the upper-pole ureter drains a single infundibulum without a true pelvis. The lower pole, which comprises two-thirds of the parenchyma, usually contains at least two major calices and a true renal pelvis.

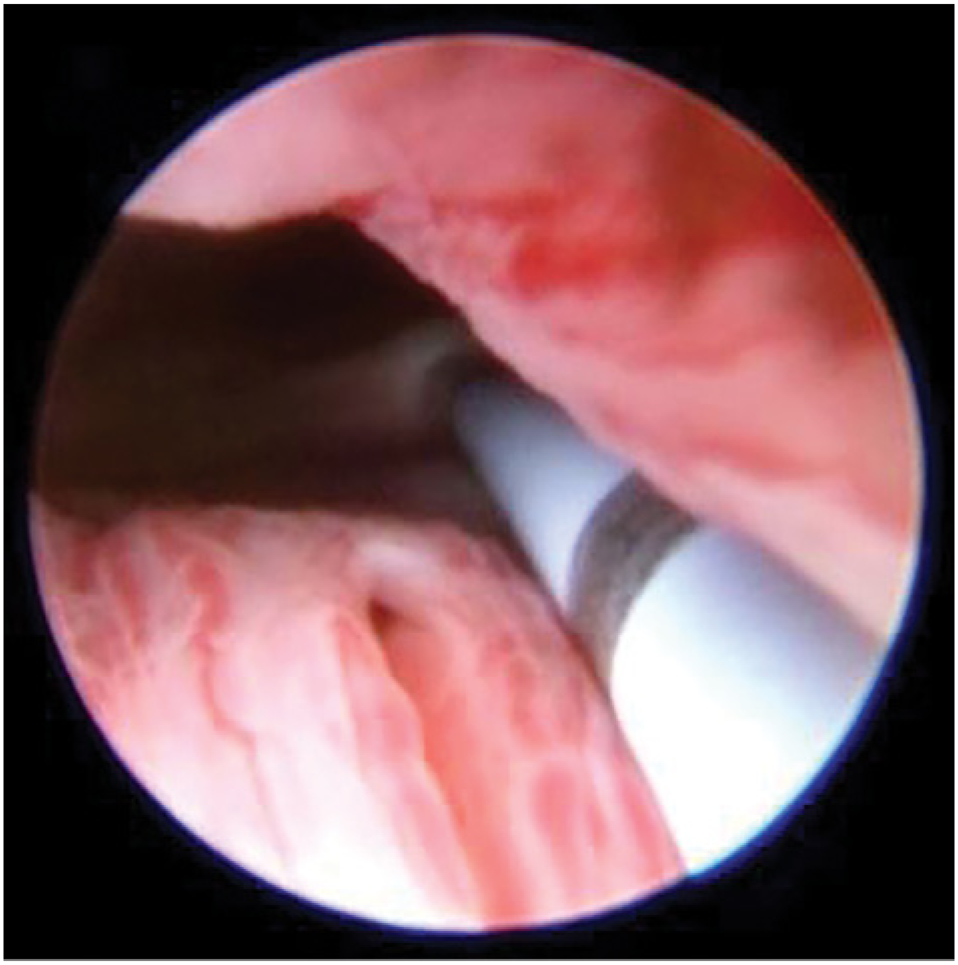


70.1 Retrograde pyelogram showing a partial duplication with obstruction of the lower ureteropelvic junction. Note the absence of a true upper renal pelvis.

Another purported complication of incomplete ureter duplication is ureteroureteral reflux, sometimes called “yo-yo” reflux. Urine that passes from one ureter to the other rather than draining antegrade is thought by some to lead to urine stasis and predispose to infection, although this is very rarely reported.

Ectopic ureter

An ectopic ureter inserts caudal to the normal location on the trigonal ridge. A ureteral bud with an abnormally high origin from the mesonephric duct (away from the urogenital sinus) may result in an ectopic ureter (**Figure 70.2**), as it incorporates into the bladder late or not at all. The ectopic orifice is always found along the pathway of the developing mesonephric system. In boys, ectopic ureters may open into bladder neck, posterior urethra (most common), or mesonephric duct derivatives: Epididymis, seminal vesicle, or vas deferens. Notably, these all sit above the urethral sphincter complex and thus never result in incontinence. By contrast, ectopic ureters in girls can terminate distal to the external sphincter and cause continuous incontinence. The most common terminal sites of ectopic ureters in girls are the bladder neck and upper urethra (33%), vaginal vestibule (33%), vagina (25%), and cervix or uterus (<5%).



70.2 An ectopic upper ureter is seen entering the bladder neck. A ureteral catheter is in the lower ureter. This patient underwent upper-to-lower ureteroureterostomy due to upper-pole obstruction.

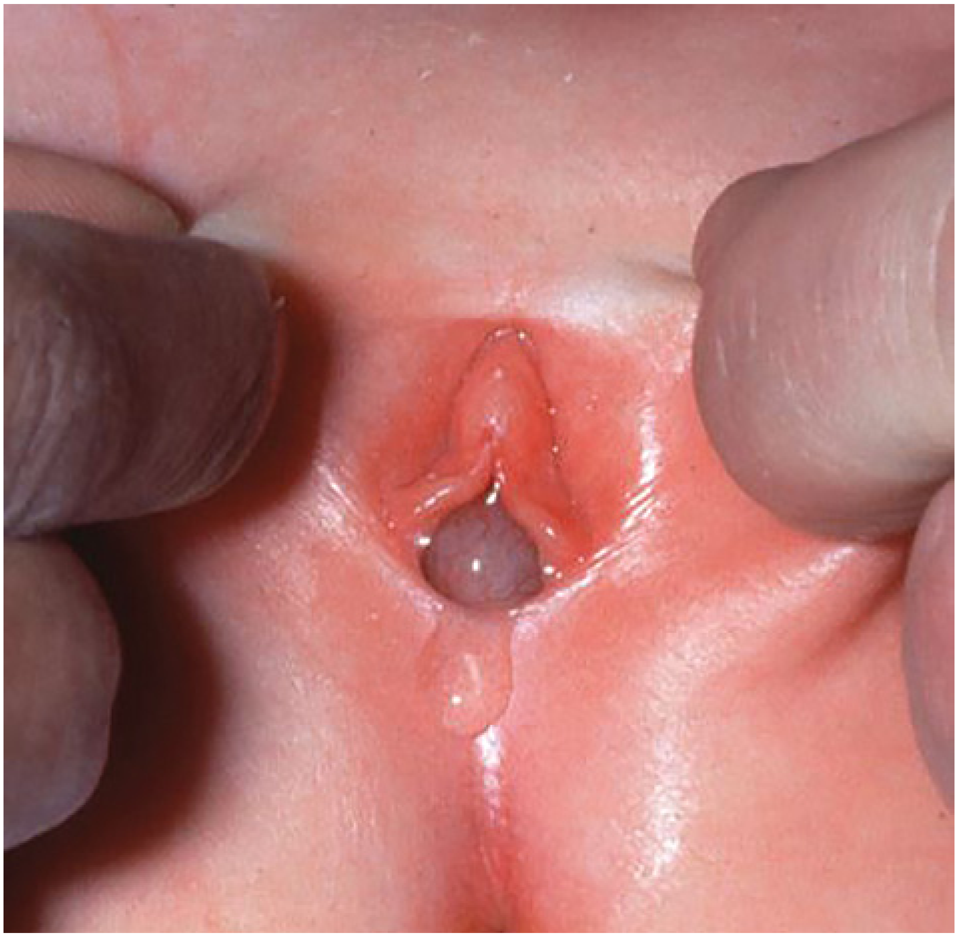
The vast majority of ectopic ureters (75–90%) are associated with the upper moiety of a duplex kidney. Nearly all ectopic ureters are associated with duplications and are found in females. When an ectopic ureter is part of a duplex system, the contralateral system is duplicated in about 80% of cases, and 20% of these have contralateral ectopy as well. Ureteral ectopia associated with a single system is comparatively rare, and has a male predominance.

Problems that result from ureteral ectopia include ureteral obstruction with resultant renal dysplasia and/or risk for infection, urinary incontinence, and occasionally epididymitis from urine reflux into the vas deferens.

Ureterocele

A ureterocele is a cystic dilatation of the terminal part of the ureter. The embryology is not entirely understood. One theory suggests that ureteroceles result from delayed canalization of Chwalla's membrane, the primitive ureteral membrane that separates the ureteric bud from the developing urogenital sinus. The distal ureteral dilation results from an obstructed or a stenotic orifice. Alternatively, some think that a ureterocele results from abnormal induction of bladder trigonal musculature.

Various classification systems have been proposed based on the location of the ureteral opening, characteristics of the orifice, or association with single or duplex collecting systems. The classification adopted by the Section on Urology of the American Academy of Pediatrics is perhaps the simplest, and categorizes ureteroceles based on the location of the dilated ureteral tissue. Ureteroceles are classified as either *intravesical* (*orthotopic*) or *extravesical* (*ectopic*). Intravesical ureteroceles are contained entirely within the bladder. Ureteroceles extending beyond the bladder neck are classified as extravesical. Occasionally, distinguishing the two can be difficult. For instance, an intravesical ureterocele may prolapse through the bladder neck (**Figure 70.3**) and an extravesical ureterocele can have an intravesical ureteral orifice. Historically, such extravesical ureteroceles with submucosal extension beyond the bladder neck were called cecoureteroceles.



70.3 Ureterocele prolapse in a female infant.

In duplex systems, ureteroceles are nearly always associated with the upper pole and 60% are extravesical. Problems include obstruction of the upper and possibly the lower pole, which in the setting of UTI can present as florid sepsis. Obstruction is typically associated with a dysplastic upper pole. Additionally, ureteroceles can prolapse and obstruct the bladder outlet. Large ureteroceles are associated with poor underlying musculature, and ureteroceles that extend beyond the bladder neck can destroy the female continence mechanism.

Vesicoureteric reflux

The combination of ureteral duplication and vesicoureteric reflux (VUR) is common. Reflux classically occurs into the lower-pole ureter, which has a laterally displaced ureteral orifice and shortened submucosal tunnel as predicted by the Weigert–Meyer law. The upper-pole ureter, with its more medial and caudal orifice, has a longer submucosal tunnel and usually does not reflux. Reflux into the upper-pole ureter may occur if its orifice is ectopic to the bladder neck or urethra, presumably because it bypasses the trigone and has an inadequate submucosal tunnel. Reflux into such ureters only occurs during voiding, and the tonically closed bladder neck may cause obstruction during the storage phase. When reflux into upper- and lower-pole ureters occurs, both orifices tend to be laterally displaced or the duplication is partial. As in non-duplicated systems, sequelae of VUR in duplicated systems include reflux nephropathy and pyelonephritis.

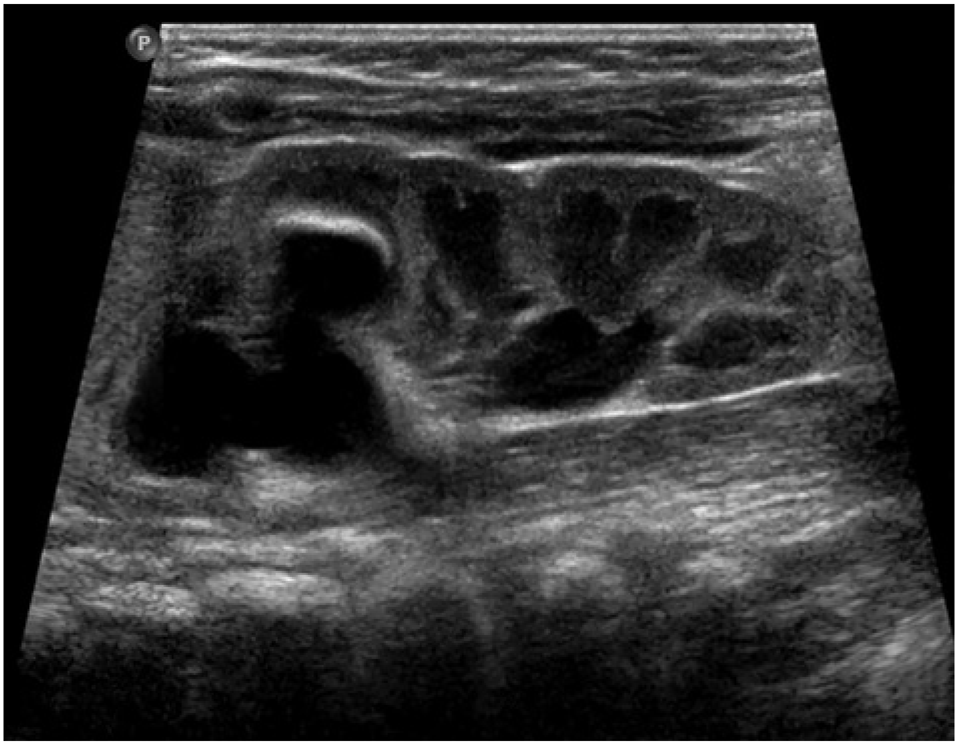
KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

The workup of ureteral duplication is targeted toward the diagnosis, prevention, and management of hydronephrosis, UTI, renal dysfunction, and voiding problems. Incidentally detected duplication in the absence of these findings does not require further investigation. Increasingly, duplication anomalies are being detected antenatally as hydronephrosis or ureteroceles are noted on obstetric sonograms. Rarely, bilateral renal dysplasia or significant renal scarring may be present, resulting in impaired renal function. In this scenario, renal function and electrolyte levels must be monitored. These patients must also be assessed for hypertension. Various tests may yield valuable information regarding complete or incomplete ureteral duplication, ectopia, ureteral dilation, ureteroceles, and relative renal function.

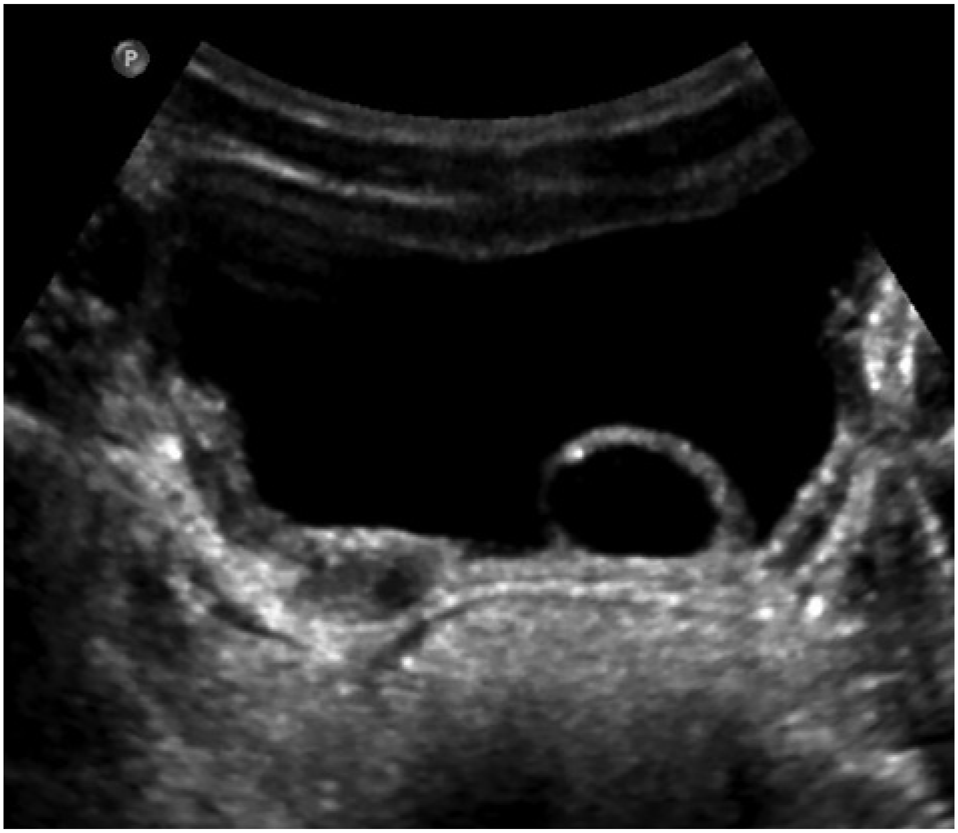
Ultrasound

Ultrasound (US) will detect upper- and/or lower-pole hydronephrosis, ureterectasis, ureteroceles, and give some information as to the function of each renal unit based on the appearance of the renal parenchyma (**Figure**

70.4). Upper hydronephrosis may occur secondary to an obstructed ectopic ureter or ureterocele. Frequently, associated hydroureter is seen. Marked dysplasia is commonly seen secondary to obstructive nephropathy. Normal renal parenchyma without ureteral dilation can suggest incidental duplication or an ectopic ureter that is not obstructed. This should prompt questions about continuous incontinence. Lower hydronephrosis is secondary to vesicoureteric or ureteroureteral reflux, UPJ obstruction, or ureteral obstruction secondary to an upper-pole ureterocele. Sonographically, a ureterocele appears as a thin-walled cystic intravesical structure (**Figure 70.5**). It may contain debris, suggestive of urine stasis or infection.



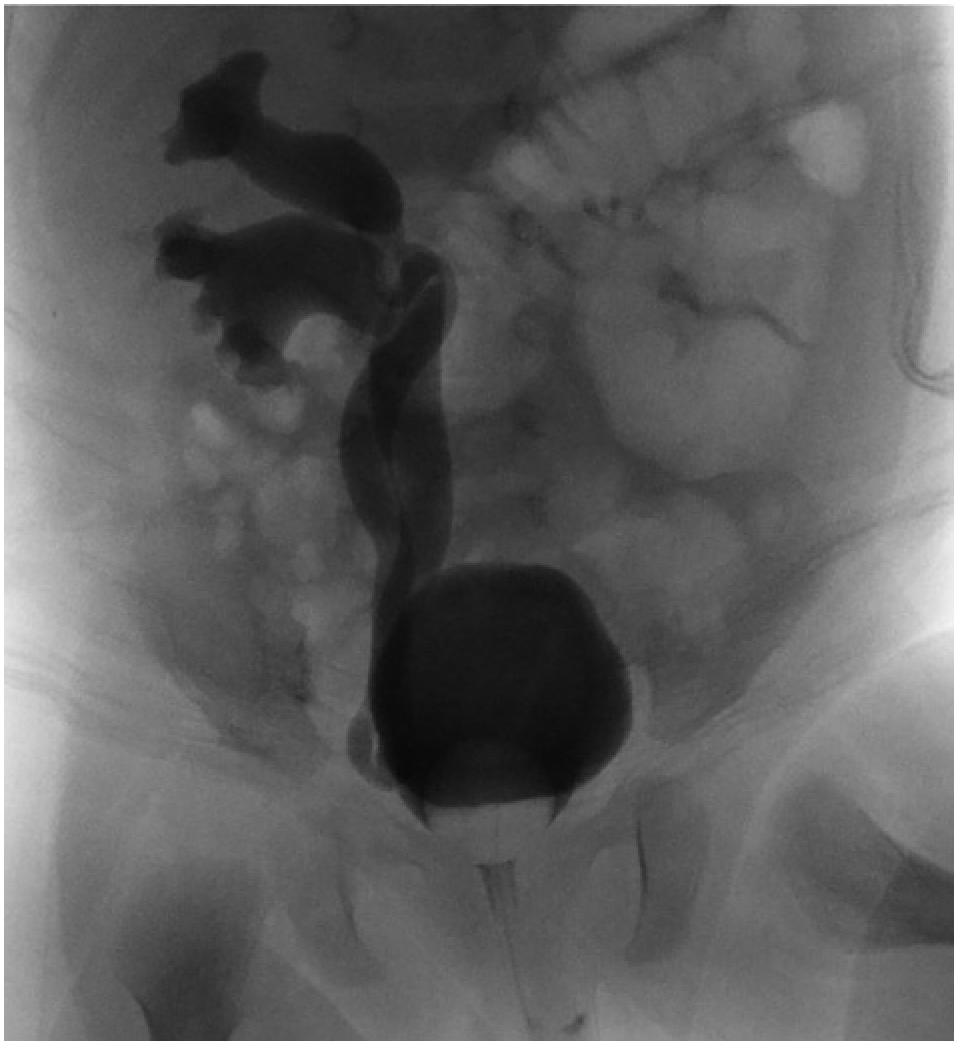
70.4 Duplex kidney with disparate hydronephrosis of the upper and lower moieties. Renal parenchyma divides the collecting systems.



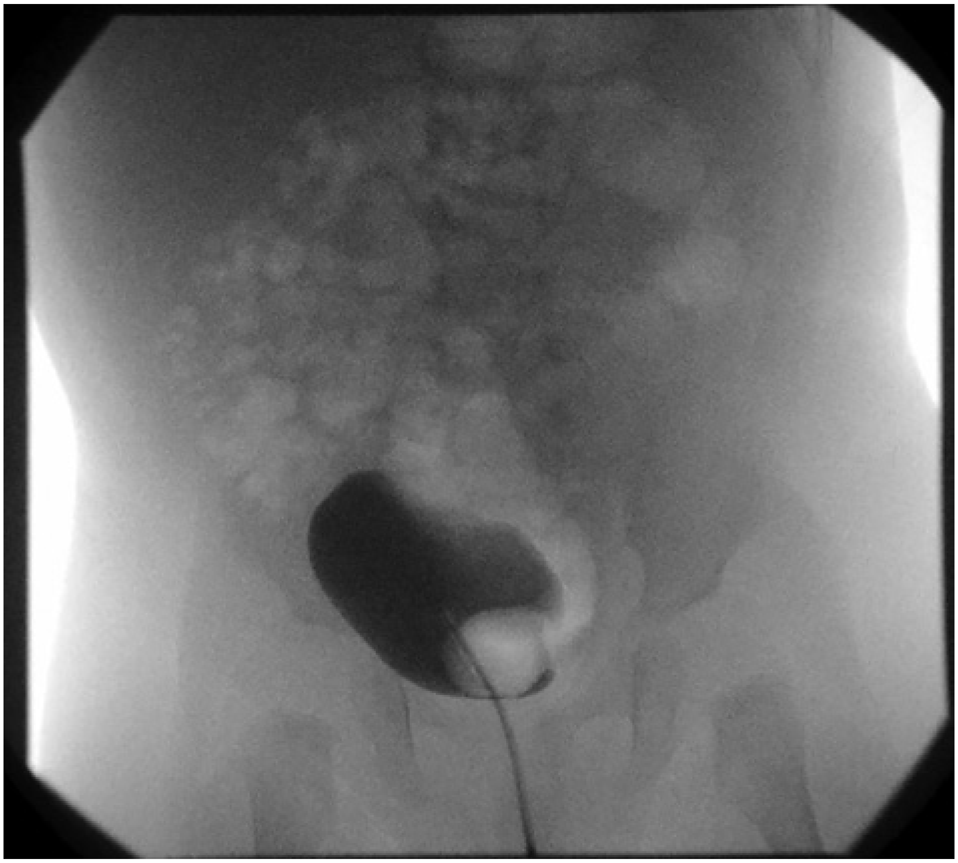
70.5 Sonographic appearance of a ureterocele.

Voiding cystourethrography

Voiding cystourethrography (VCUG) is the study of choice to detect and characterize VUR. In the present of reflux, missing calyces or a “drooping lily sign” suggests duplication (**Figure 70.6**). VCUG also may show ureteroceles, which are best seen during early filling (**Figure 70.7**). The dynamic nature of this modality yields information regarding bladder neck obstruction, ureterocele prolapse, ureterocele eversion (which suggests poor trigonal musculature), and ureteral dilation.



70.6 Vesicoureteric reflux into both moieties of a duplex system. The catheter balloon is inflated; this should not be mistaken for a ureterocele.



70.7 A ureterocele is visible as a filling defect in the bladder early in the filling phase of a voiding cystourethrogram. As filling progresses, the ureterocele is obscured, compressed, or can evert and become difficult to identify.

Renal scintigraphy

Radio isotope renal scans show relative function of the renal units and may reveal a "cryptic" duplex kidney. In the non-pathologic duplex kidney, the relative function of the upper and lower moieties is one-third and two-thirds, respectively.

Magnetic resonance urography

Magnetic resonance urography (MRU) provides precise anatomic detail in patients with complex anatomy that may be difficult to visualize with other modalities. MRU may be especially useful in tracing ectopic ureters. MRU is also useful in patients without ureteral dilation, in those with ureteroceles that obscure other aspects of the bladder, and in patients with uterine, vaginal, or other bladder anomalies. There is now available software, albeit expensive and complex, which allows computation of differential renal function and urine transit times. This may eventually obviate the need for renal scintigraphy.

Intravenous urography

With other modalities available, intravenous urography (IVU) is now rarely performed. It can reveal duplication and the level of confluence, ureteral ectopia, and ureteroceles. When upper moiety function is poor, associated pathology is not always shown and delayed images may be helpful.

Cystoscopy

Direct endoscopic evaluation is useful to characterize a ureterocele and/or locate the ureteral orifice. If deemed useful, vaginoscopy and/or retrograde urography can be performed simultaneously.

TREATMENT

Medical treatment has a limited role in the management of duplication anomalies. Antibiotic prophylaxis is often prescribed for patients with ureteral obstruction and/or high-grade VUR, or patients with a history of UTI. Treatment of any underlying bowel–bladder dysfunction is key to the management of VUR, as it is in single systems. Thereafter, treatment of ureteral duplication anomalies is surgical.

There are now endoscopic, laparoscopic/robotic, and open surgical techniques to correct the many clinical problems presented by ureteral duplication. Choosing the correct approach requires a thorough understanding of the underlying disease. Frequently, patients will require two operations–one temporizing procedure, followed by definitive reconstruction when the patient is older, larger, and medically stable (**Table 70.2**).

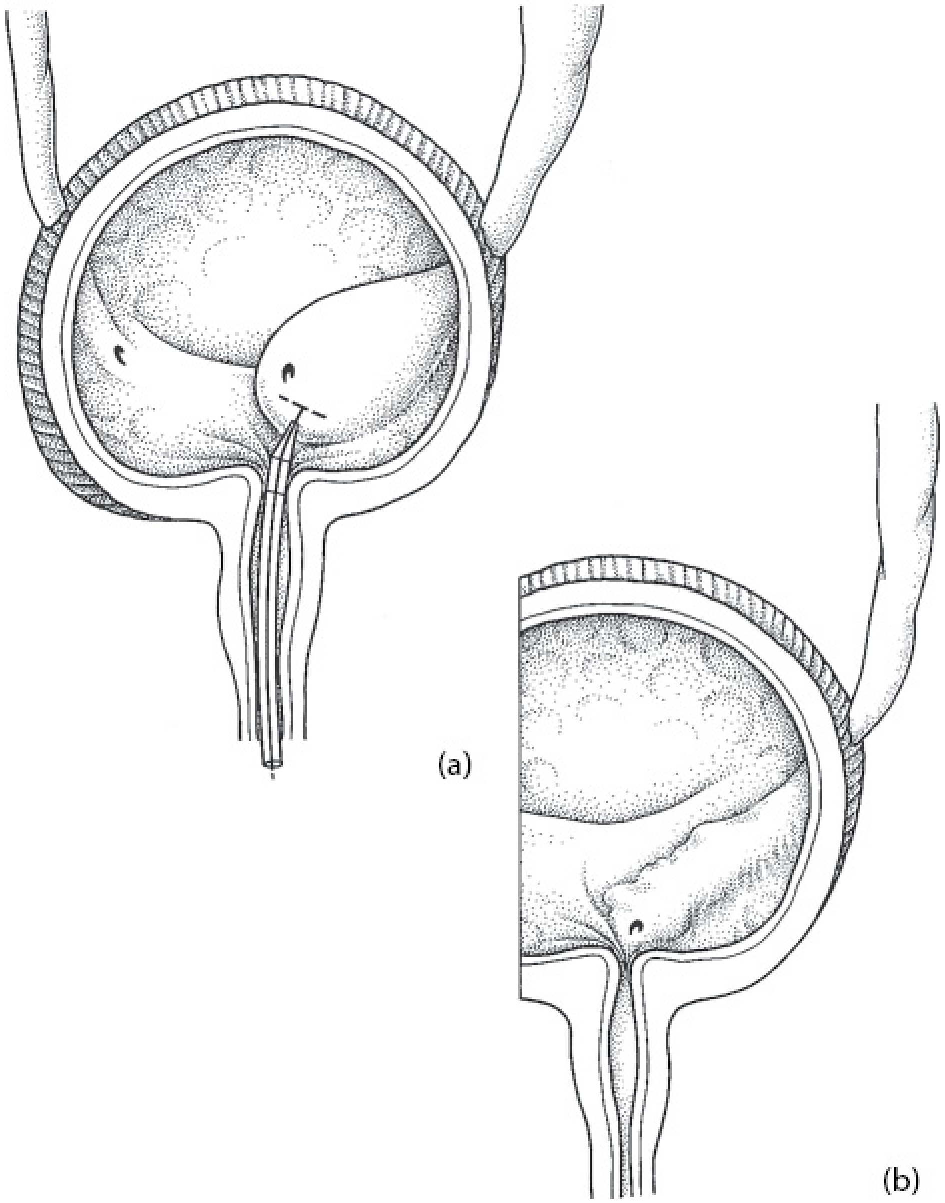
Table 70.2 **Definitive reconstructive options for the typical duplex system**

	Functional upper pole	Non-functional upper pole
Bladder pathology	Ureteral reimplantation	Ureteral reimplantation
No bladder pathology	Ureteroureterostomy (high or low)	Heminephrectomy Ureteroureterostomy (high or low)

ENDOSCOPIC OPERATIONS

Ureterocele incision

Endoscopic decompression of the ureterocele was initially suggested in 1962 by Zielinski. The initial approach was to unroof the ureterocele, which was successful but incurred a high rate of de novo VUR. Currently, the more common approach is a low, medial, transverse incision at the base of the ureterocele (**Figure 70.8a**). This provides adequate ureteral decompression while residual ureterocele tissue creates a functional flap valve to prevent VUR (**Figure 70.8b**). The opening created should be intravesical to avoid obstruction by the bladder neck. The incision can be completed with electrocautery, laser, or a cold knife. Successful initial decompression is seen in upward of 90% of patients.



70.8a,b

More recently, a “watering can” approach has been advocated in the absence of infection wherein the ureterocele is punctured 10–20 times with a laser. This technique yields durable decompression with markedly reduced iatrogenic reflux, however the need for secondary procedures is unknown.

For intravesical ureteroceles, regardless of technique, primary endoscopic decompression is frequently the only intervention needed. For ectopic ureteroceles, the long-term outcomes are not good. Ureterocele decompression can be viewed as a temporizing operation, as 50–80% of patients will require secondary reconstruction.

Subureteric injection

As in single systems, VUR in duplex systems can be managed with subureteric injection of bulking agents. Success is lower than in single systems and is related to age, sex, and degree of reflux. Multiple injections do increase resolution of reflux, albeit with diminishing returns. Long-term success rates fall to well under 50% with high-grade reflux, particularly when associated with bladder dysfunction, bilaterality, younger age, and male gender. Late failures are thought to be due to migration of the bulking agent, which may be more common in duplex ureters due to poor detrusor backing. Ureteral obstruction is rare but has been reported, and can lead to renal loss. The manufacturer of dextranomer/hyaluronic acid (Deflux® [Palette Life Sciences, Stockholm, Sweden]) has recently removed duplication as a contraindication.

BLADDER-LEVEL (LOWER TRACT) RECONSTRUCTION

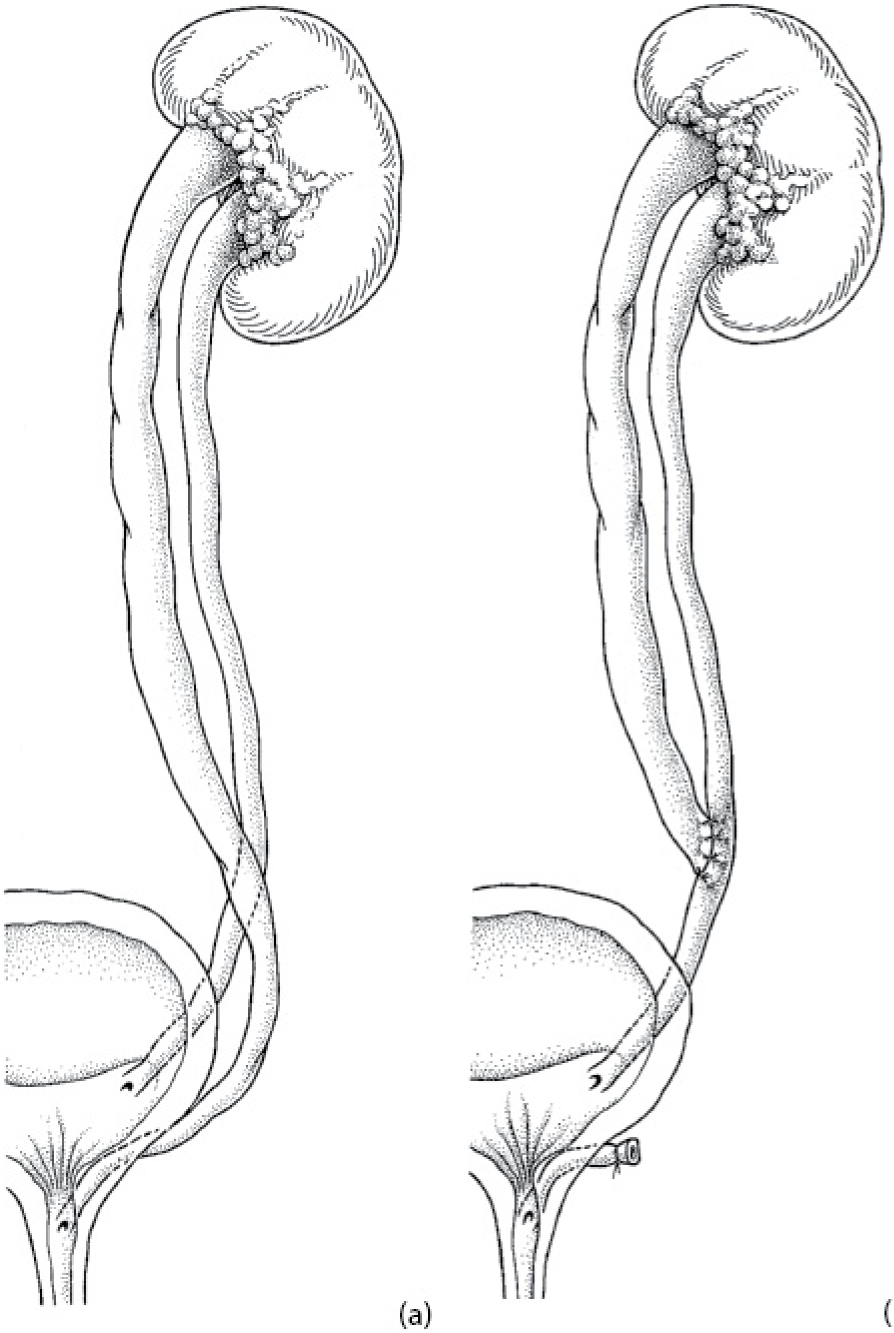
Definitive urinary tract reconstruction can occur at the bladder level (lower tract) or at the kidney level (upper tract). Surgeons should try to avoid combined upper and lower tract approaches, as they are more invasive and rarely necessary. If a patient has bladder pathology including VUR or a large or ectopic ureterocele, a lower tract approach is best.

Open extravesical operations (e.g. ureteroureterostomy) are conducted through an inguinal or Gibson incision, depending on patient age. The floor of the inguinal canal is opened, the perivesical space is developed and the ureters are found posterolateral to the medial umbilical ligament. Some will pre-place ureteral stents in non-dilated ureters to aid in their identification. In males, care is taken to avoid damaging the vas deferens, which drapes over the ureters. Intravesical operations (e.g. cross-trigonal ureteral reimplantation) are performed through a Pfannenstiel incision.

Laparoscopic or robotic approaches involve traditional port placement for pelvic surgery, including an umbilical camera port and left and right infraumbilical working ports.

Ureteroureterostomy

Ureteroureterostomy is an excellent solution when there is isolated upper- or lower-pole pathology, but not both. If there is isolated lower VUR, the lower ureter can be anastomosed to the upper ureter in an end-to-side fashion. More commonly, an upper-to-lower ureteroureterostomy is performed in the setting of an ectopic ureter causing urinary incontinence or obstruction (**Figure 70.9a and b**). Upper-pole ureters that are obstructed by small intravesical ureteroceles may also be managed with ureteroureterostomy, however large ureteroceles or ectopic ureteroceles frequently require an intravesical operation for formal ureterocele excision and bladder floor reconstruction. If there is no ureteral dilation, the ureter should be spatulated to allow a widely patent anastomosis. The distal upper ureteral stump is not aggressively pursued so as not to damage the shared blood supply with the distal lower ureter.



70.9a,b

Risks include ureteroureteral reflux, damage to the recipient ureter, anastomotic stenosis, urine leak, and infection of the ureteral stump, all of which are rare. If the ureters share a common sheath, excision of the distal ureteral stump should not be aggressively pursued in order to avoid damaging the shared blood supply. Stents, if placed, can either traverse the anastomosis or remain in the recipient ureter. Success is defined as improved or resolved hydronephrosis on postoperative US, and exceeds 90% in experienced hands.

Ureteral reimplantation

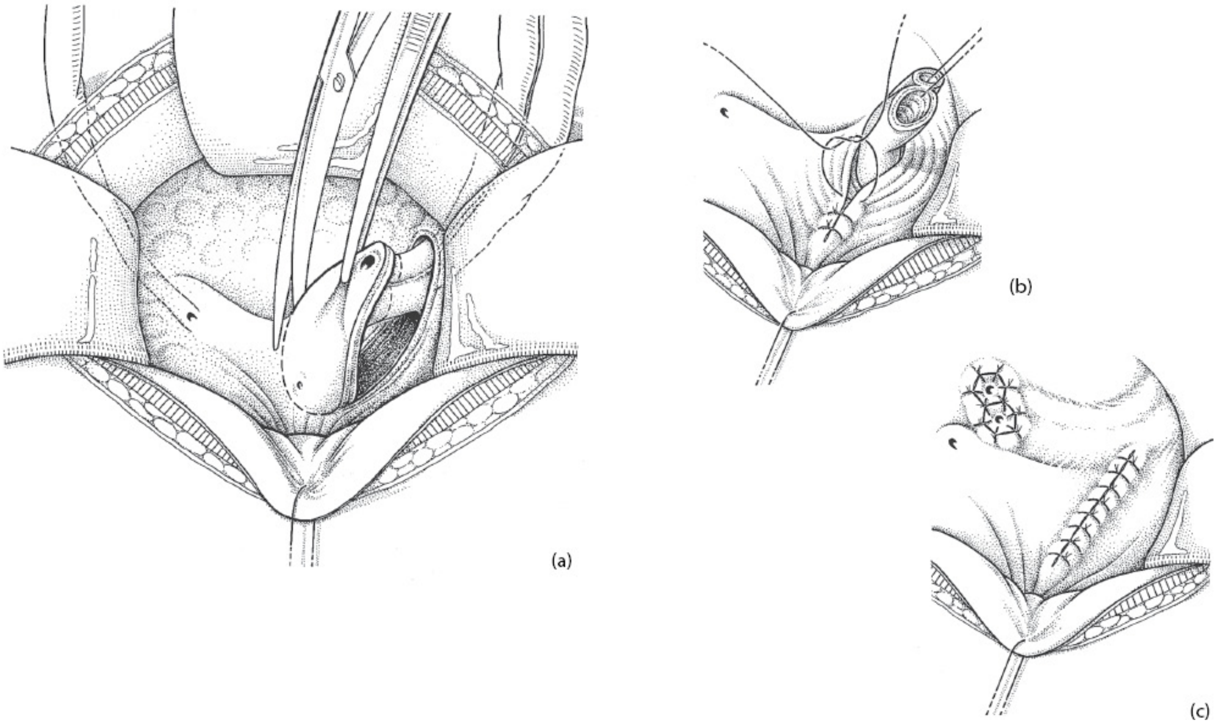
Isolated VUR in duplex systems is treated as it is in single systems. When VUR is present in addition to an ectopic or obstructed ureter requiring intervention, ureteral reimplantation is the procedure of choice. Distally, duplicated ureters are often located within a common sheath and share a distal blood supply. In this case, ureters must be mobilized and reimplanted en bloc. An ectopic ureter will not necessarily share a blood supply with the lower-pole ureter. In this case, the ureters can be reimplanted independently, although this is technically very difficult and it is often better to excise the distal part of the ectopic ureter and perform an en bloc ureteric reimplant of both ureters. Alternatively, a combination upper-to-lower ureteroureterostomy and lower ureteral reimplant may be performed.

Ureters may be reimplanted via an intravesical or extravesical approach, with or without ureteral tailoring. Bilateral extravesical reimplantation is avoided due to the risk of pelvic plexus damage and urinary retention. Reimplantation of obstructed or ectopic ureters, ureteral tailoring, and excision of associated ureteroceles requires dismembering the ureter and opening the bladder mucosa, thus many surgeons prefer an intravesical approach in this setting.

While laparoscopic intravesical ureteral reimplantation has been described, the traditional laparoscopic approach is extravesical. The success rate of common sheath reimplantation is >95%, similar to single-system reimplantation.

INTRAVESICAL APPROACH

A Pfannenstiel incision is made, and the bladder is opened and retracted cephalad to expose the trigone (**Figure 70.10a**). Ureteral catheters are inserted. The common sheath double ureters are mobilized and brought through a cross-trigonal submucosal tunnel that is five times the length of the ureteral lumen (**Figure 70.10b**). The detrusor defect of the native hiatus is closed and the ureters are tunneled together submucosally, taking care not to damage the contralateral ureter (**Figure 70.10c**). Stents are unnecessary in the absence of ureteral tailoring. The bladder is drained overnight.



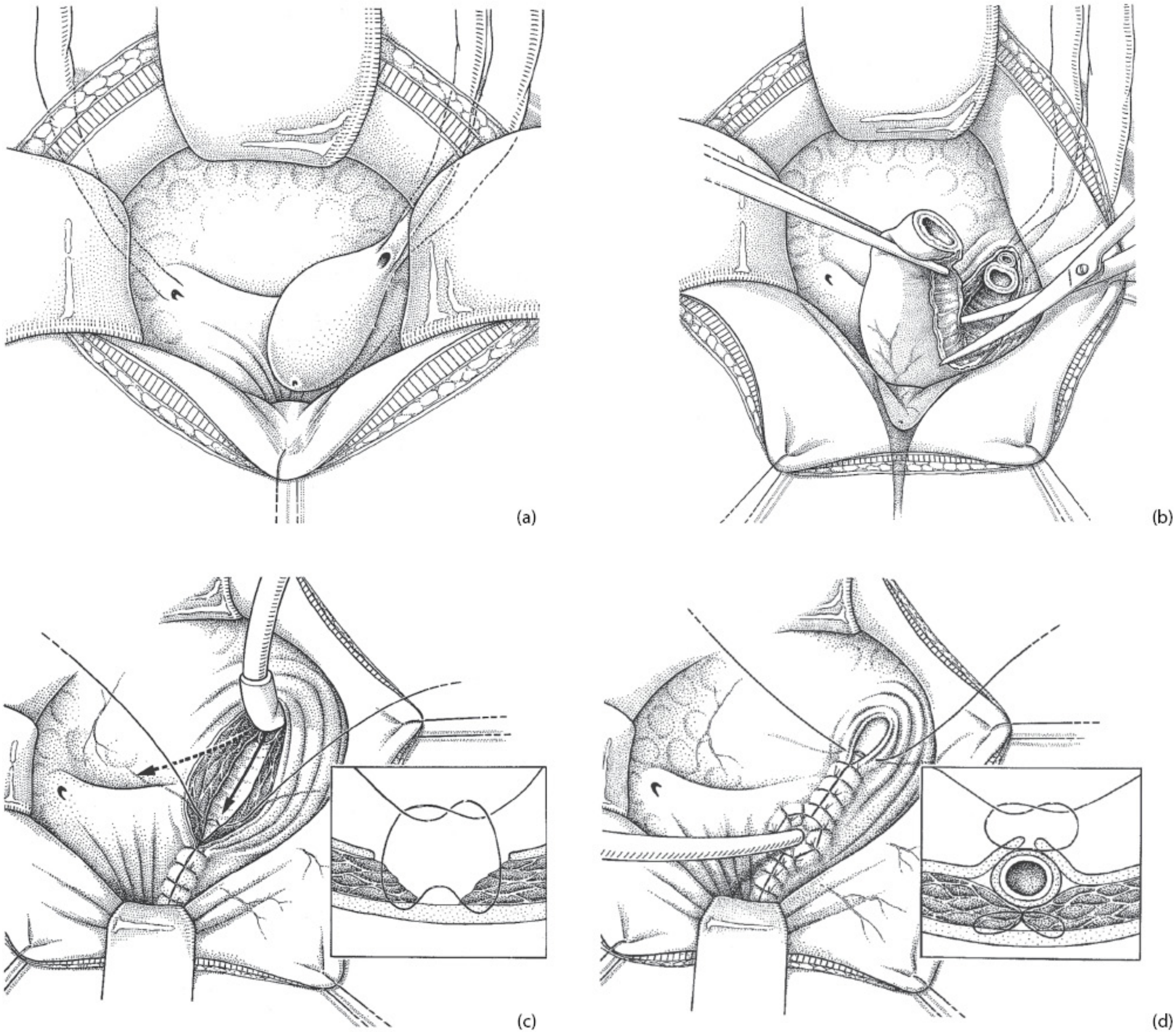
70.10a–c

EXTRAVESICAL APPROACH

The open approach is via an inguinal, Gibson, or Pfannenstiel incision. Laparoscopic ports are configured for pelvic surgery. A catheter is placed on the field to control distension of the bladder. The obliterated umbilical artery is retracted medially revealing the ureters, which are mobilized to the vesicoureteric junction (VUJ). An extravesical detrusorotomy is made from the VUJ toward the contralateral shoulder to avoid ureteral kinking when bladder traction is released. The detrusor is closed around the inlaid ureters. Routine catheter drainage is not required in the absence of a cystotomy.

EXCISION OF URETEROCELE

Excision of the ureterocele is performed at the time of ureteral reimplantation (**Figure 70.11a and b**). Redundant mucosa is excised, taking care not to leave a mucosal flap which can obstruct the bladder. The detrusor defect is reconstructed to ensure adequate backing for the reimplanted ureter. The ureter is laid over this (**Figure 70.11c**) and the overlying mucosa is closed, recreating the submucosal tunnel (**Figure 70.11d**). The upper ureter is not shown but is typically reimplanted along with the lower ureter.



70.11a–d

Contralateral reflux can result, presumably due to trigonal distortion, though the clinical significance is unclear. When the ureterocele extends past the bladder neck, there is a significant risk of stress urinary incontinence and bladder dysfunction. Prudent dissection is required, but even with meticulous technique, outcomes are poor. Patients may require formal bladder neck reconstruction or additional outlet procedures.

There is no consensus as to the best approach to the extravesical ureterocele. Rates of bladder dysfunction and urinary incontinence are similar regardless of the surgical strategy, including upper-tract only intervention, suggesting that these may be congenital rather than iatrogenic problems. Outcomes in the literature are difficult to interpret due to small series, changing classifications of ureteroceles, varied surgical approaches, inconsistent indications for secondary surgery, and vague definitions for bladder dysfunction and surgical success. Bladder dysfunction in these patients is thought to be due to myogenic failure and responds well to behavioral and medical therapy but is unaffected by surgical intervention. By contrast, stress urinary incontinence is difficult to treat, and is refractory even to bladder neck reconstruction. Small series from experienced surgeons show that 50–100% of patients required placement of an artificial urinary sphincter or bladder-neck closure after bladder-neck reconstruction to address persistent incontinence.

Cutaneous ureterostomy

This is a temporizing operation performed in the setting of UTI in the young patient. It is indicated for the infected, obstructed upper-pole ureter or the refluxing lower-pole ureter with breakthrough infections on antibiotic prophylaxis. The surgical approach is identical to a ureteroureterostomy. The pathologic ureter(s) are incised or transected and marsupialized to the inguinal skin incision. In infants, the combination of small bladders and dilated ureters precludes an adequate ureteral tunnel, and reimplantation is best delayed. Ureteral dilation often improves with diversion, which may obviate the need for tailoring at the time of reimplantation.

Ureteroureterostomy or ureteral reimplantation can be undertaken when the patient is 1–2 years of age. Notably, diverting ureterostomy is an excellent option for managing ureteroceles when endoscopic equipment is not available.

KIDNEY-LEVEL (UPPER TRACT) RECONSTRUCTION

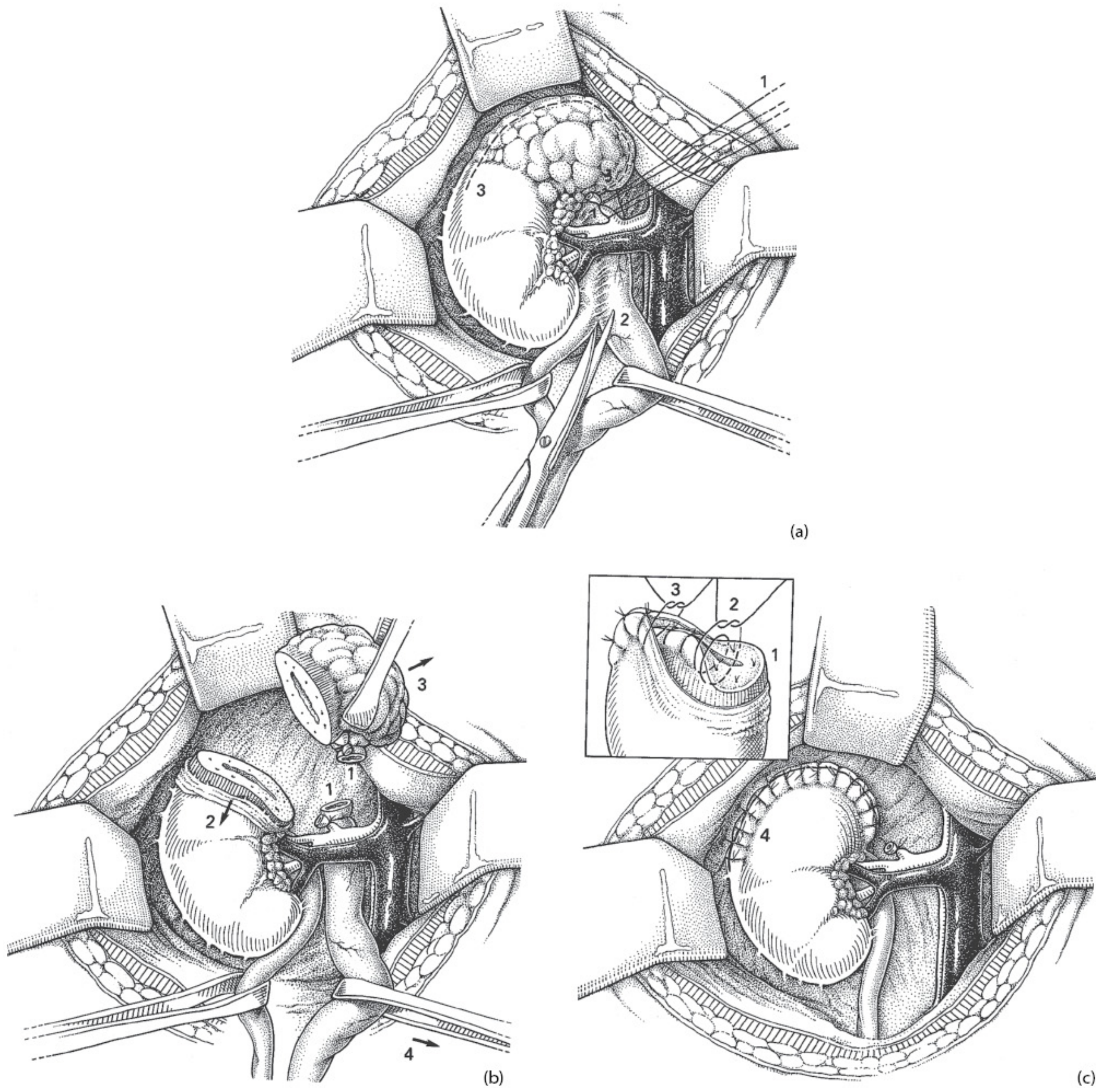
Upper tract reconstruction is appropriate only when there is no lower urinary tract pathology to be addressed. Even so, a lower tract approach is often considered due to its relative ease and success.

The open approach to the kidney is through a retroperitoneal flank incision. The patient is positioned in the modified lateral decubitus position and an incision is made from the tip of the twelfth rib toward the umbilicus. The muscles are split and the retroperitoneum is entered off the tip of the twelfth rib. The peritoneum is swept medially. Gerota's fascia is entered, exposing the kidney and collecting system.

The laparoscopic or robotic approach to urinary tract reconstruction is transperitoneal. The patient is similarly positioned, and an umbilical camera port is placed along with two triangulating working ports, one in the ipsilateral lower quadrant and the other in the upper midline or ipsilateral subcostal midclavicular line. Additional retraction ports are sometimes placed. The colon is reflected medially, exposing the retroperitoneum. Transabdominal stay sutures through the collecting system are often helpful.

Heminephrectomy

The indication for upper heminephrectomy (**Figure 70.12a–c**) is a poorly functioning upper pole with associated urinary incontinence or upper-pole pathology (e.g. pyelonephritis, stones). The upper-pole ureter is divided and used for traction. The upper-pole hilar vessels are ligated, and the non-viable renal parenchyma demarcated. The upper-pole parenchyma is excised and closed in layers as a renorrhaphy. These sutures must pass through the renal capsule to avoid tearing through parenchymal tissue.



70.12a–c

Alternatively, the upper-pole parenchyma, which is often poorly vascularized and easily delineated from the healthy lower pole, can be excised and traced to the upper-pole hilar vessels and ureter, which are then divided. This simplifies the hilar dissection but risks bleeding. The cut edge of renal parenchyma is covered with Gerota's fascia and perirenal fat. Redundant upper-pole ureter is removed but the residual ureteric stump is left *in situ*. Utmost care is taken not to damage the blood supply to the lower-pole parenchyma or ureter.

Loss of lower-pole function and/or conversion to total nephrectomy is the most significant complication related to heminephrectomy, with reported rates as high as 10%. This can be due to hilar injury or postoperative torsion of the remaining kidney and is more common in infants. For fully mobilized kidneys, consider nephropexy to prevent torsion of the renal remnant. Additional complications include postoperative bleeding (<5%), urinoma (5–20%, more common in laparoscopic procedures, rarely clinically significant) and damage to adjacent vascular and visceral structures (rare).

In the absence of lower-pole pathology, fewer than one in ten patients will require further surgery. In the presence of an ectopic ureterocele or VUR, heminephrectomy is a poor choice as further operation is required in well over half of children. Common secondary operations include ureteral reimplantation, excision of the ureteral stump, and ureterocele excision. Notably, these data arise from an era of aggressive surgical treatment of VUR; reoperation rates may be markedly lower today. While upper heminephrectomy was once the procedure of choice for the non-functional upper pole without lower-pole reflux, practice patterns are shifting toward ureteroureterostomy, which is a simpler operation with similar success rates and less risk to lower-pole function.

Lower heminephrectomy is uncommon. The indication is a non-functional lower renal unit secondary to UPJ obstruction or reflux nephropathy with preservation of the upper pole. Technique, complications, and outcomes are similar to upper-pole heminephrectomy.

Pyelopyelostomy/ureteropyelostomy

An upper-to-lower pyelopyelostomy or ureteropyelostomy can be considered when there is upper-pole ureteral obstruction secondary to an ectopic ureter or ureterocele. The advantage over a ureteroureterostomy is that the recipient lower-pole pelvis is larger caliber than the ureter, and thus there is less risk of damage to the recipient drainage system. In this operation, the lower-pole pelvis is incised, the upper-pole ureter is transected, and the two are anastomosed in an end-to-side fashion. The distal upper-pole ureteric stump can be aspirated and/or partially excised to minimize urine stasis.

A lower-pole UPJ obstruction is an uncommon finding in incomplete ureteral duplication. Typically the lower-pole ureter is short, precluding a traditional pyeloplasty. The upper-pole collecting system is divided and the lower renal pelvis is opened. A side-to-side anastomosis between the obstructed lower-pole renal pelvis and the upper-pole ureter is performed. Care must be taken at the apices of the anastomosis as the upper-pole ureter is easily narrowed. Most surgeons will place an indwelling ureteral stent in the recipient (upper-pole) ureter. In the presence of an internal stent, periureteral drain placement is unnecessary. Complications and outcomes are similar to ureteroureterostomy.

Nephrectomy

Nephrectomy is indicated for a problematic non-functional kidney, and is described elsewhere (see Chapter 68).

POSTOPERATIVE CARE

Most operations can be performed on an outpatient basis or with overnight observation. Dietary restrictions are unnecessary. No stents or drains are needed for nephrectomies. Many surgeons elect to place ureteral stents or drains following ureteral reconstruction. Overnight bladder drainage is appropriate following operations in which the bladder is opened. Renal US should be obtained 6–12 weeks postoperatively to ensure renal decompression.

FURTHER READING

Castagnetti M, El-Ghoneimi A. Management of duplex system ureteroceles in neonates and infants. *Nat Rev Urol* 2009; 6: 307–15.

Nepple GK, Cooper CS, Snyder HM. Ureteral duplication, ectopy and ureteroceles. In: Gearhart J, Rink R, Mouriquand P (eds). *Pediatric Urology*, 2nd ed. Philadelphia: Saunders Elsevier; 2010: pp. 337–52.

Peters CA, Schlussel RN, Mendelsohn C. Ectopic ureter, ureterocele, and ureteral anomalies. In: Wein AJ, Kavoussi LR, Partin AW, Peters CA (eds). *Campbell-Walsh Urology*, 11th ed. Philadelphia: Saunders Elsevier; 2016: pp. 3075–101.

Rickwood AMK, Madden NP, Boddy SM. Duplication anomalies, ureteroceles and ectopic ureters. In: Thomas DFM, Rickwood AMK, Duffy PD (eds). *Essentials of Paediatric Urology*, 2nd ed. London: Martin Dunitz; 2008: pp. 93–108.

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Urinary diversion

PRASAD GODBOLE

PRINCIPLES AND JUSTIFICATION

Urinary diversion, although uncommon in children, can be a useful tool to a pediatric urologist. It is often used for short-term drainage of the urinary tract postoperatively for a variety of surgical procedures (e.g. anorectal malformation, posterior urethral valves (PUVs), stone surgery). The primary aim is to ensure adequate drainage of the urinary tract and thereby preserve renal function. The secondary aim is to achieve continence. This chapter will not focus on the routine procedures requiring urinary diversion such as urethral/suprapubic bladder drainage or nephrostomy but will primarily cover longer-term or permanent urinary diversion.

Diversions may be incontinent or continent. Incontinent diversions entail cutaneous drainage of urine into a pad, diaper, or stoma bag. A continent diversion aims to create a low-pressure, compliant, and capacious reservoir and a catheterizable conduit for drainage of the reservoir where the urethra is found to be unsuitable.

The majority of urinary diversions are performed for congenital anomalies of the urinary tract and their consequences. Examples include neuropathic bladder due to spinal dysraphism, the exstrophy epispadias complex, PUVs, functional bladder disorders such as the Hinman bladder, or bladder malignancy. The choice of diversion depends on the child's diagnosis, body habitus, manual dexterity, compliance, and social circumstances. For example, a child with a neuropathic bladder, severe developmental delay, difficult social circumstances, and deteriorating upper tracts may be better off with a vesicostomy rather than a bladder augmentation and a Mitrofanoff conduit.

PREOPERATIVE ASSESSMENT AND PREPARATION

The assessment and preparation of children for surgery for urinary diversion consist of three components:

- *Overall assessment*, e.g. severe kyphoscoliosis may need respiratory preparation.
- *Renal assessment*, e.g. chronic renal requires nephrology input. Ultrasound scan and functional imaging – either a mercaptoacetyl triglycine (MAG3) or dimercaptosuccinic acid (DMSA) scan; additional investigations depend on individual patients, e.g. videourodynamics, micturating cystourethrogram.
- *Assessment for the specific procedure*

Assessment and preparation for the specific procedure

The capabilities, compliance, and circumstances of the child and carers must be taken into consideration before a decision can be made. This is conducted in part by clinicians, but also by a team of clinical nurse specialists. Preparation is augmented by information leaflets and other visual aids. The child may also be encouraged to interact with other children who have had a similar procedure.

Bowel preparation may be needed, particularly if the colon is being used (e.g. colocystoplasty) but this need be no more restrictive than clear fluids for 24 hours preoperatively. In general, for most urological procedures, broad-spectrum antibiotics such as amikacin alone or a combination of amikacin, benzylpenicillin, and metronidazole, at induction and for up to 48 hours postoperatively are used.

Urinary diversions can be divided accordingly:

- Incontinent diversions
  - Open or button vesicostomy; ureterostomy and pyelostomy
  - *Conduit diversion*: Ileal conduit, colonic conduit
- Continent diversions
  - *Surgery of the reservoir*: Ileocystoplasty, colocystoplasty
  - *Continent catheterizable conduit*: Mitrofanoff principle and its variants: Appendicovesicostomy, ileovesicostomy (Yang–Monti), detrusor tube vesicostomy
  - *Stoma*: VQZ plasty

INCONTINENT URINARY DIVERSION

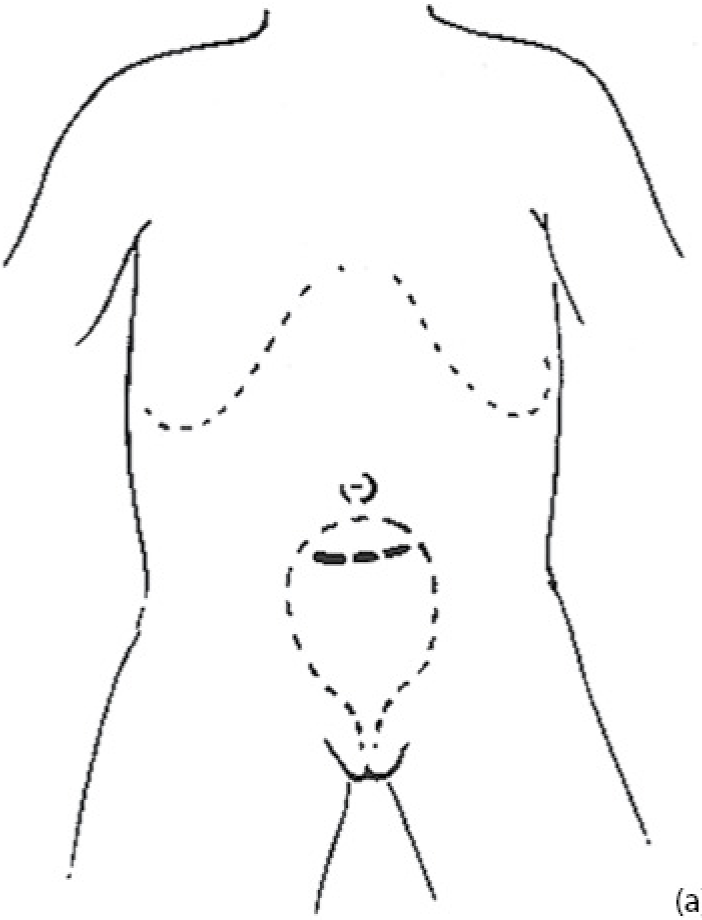
Vesicostomy

A vesicostomy may be performed as a temporary measure for urinary diversion in selected cases such as PUV, gross vesicoureteric reflux (VUR), or neuropathic bladder. Although primary fulguration is the ideal method of treatment in cases of PUV, we have found there to be no difference in the outcome for boys who had an initial vesicostomy and delayed ablation. Vesicostomy does have a high incidence of stomal stenosis and prolapse in the long term and should not be considered as a permanent option. Furthermore, care of the skin around the vesicostomy is important as quite often it may become excoriated due to contact with urine.

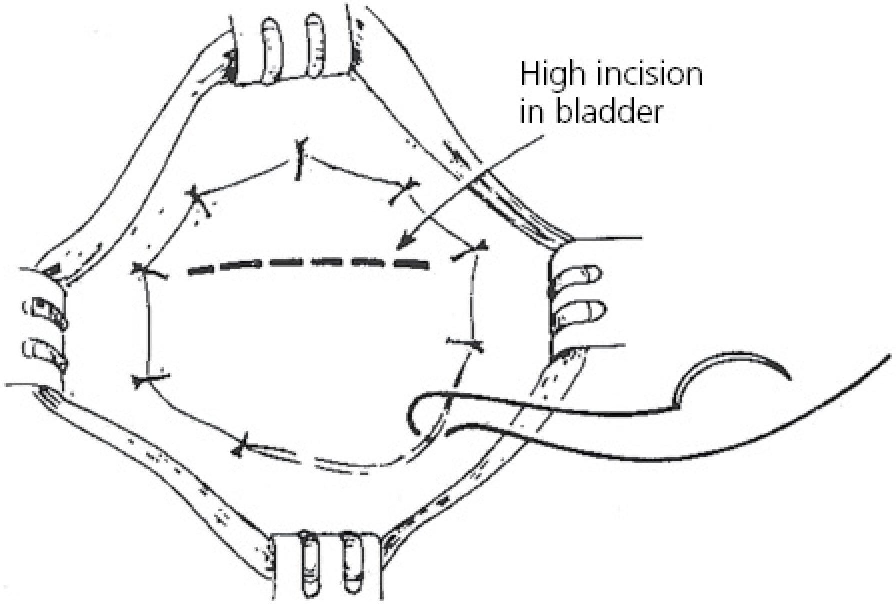
An alternative to vesicostomy is the use of a suprapubic gastrostomy button inserted via an open technique primarily or inserted after initial suprapubic catheter drainage utilizing the same tract. This allows intermittent drainage of urine in a “continent” manner.

TECHNIQUE

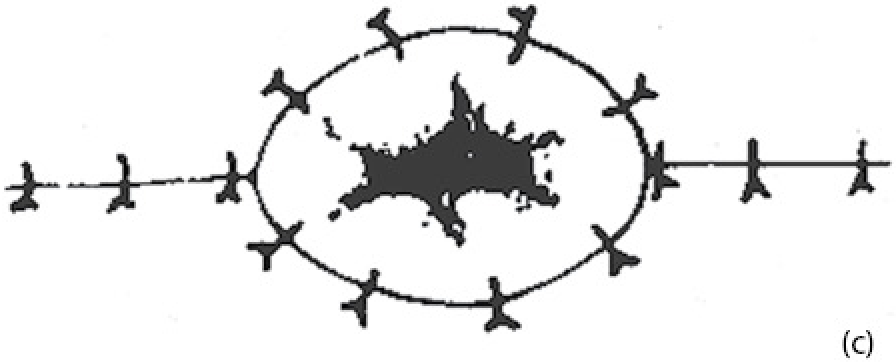
The patient is placed in the supine position. A short transverse incision is made above the pubic symphysis in an area that will drain into the pad/diaper (**Figure 71.1a**). The bladder is mobilized extraperitoneally and the dome is identified by the urachal remnant, which is ligated with an absorbable suture (**Figure 71.1b**). The dome is sutured circumferentially with interrupted 4/0 absorbable sutures to the fascia. The dome is then opened and the mucocutaneous anastomosis is performed with circumferential 4/0 absorbable, interrupted sutures (**Figure 71.1c**).



(a)


High incision
in bladder

(b)



71.1a–c

A 12 Fr catheter is left *in situ* for 24 hours and then removed. Subsequently, the vesicostomy is allowed to drain into the diaper.

COMPLICATIONS

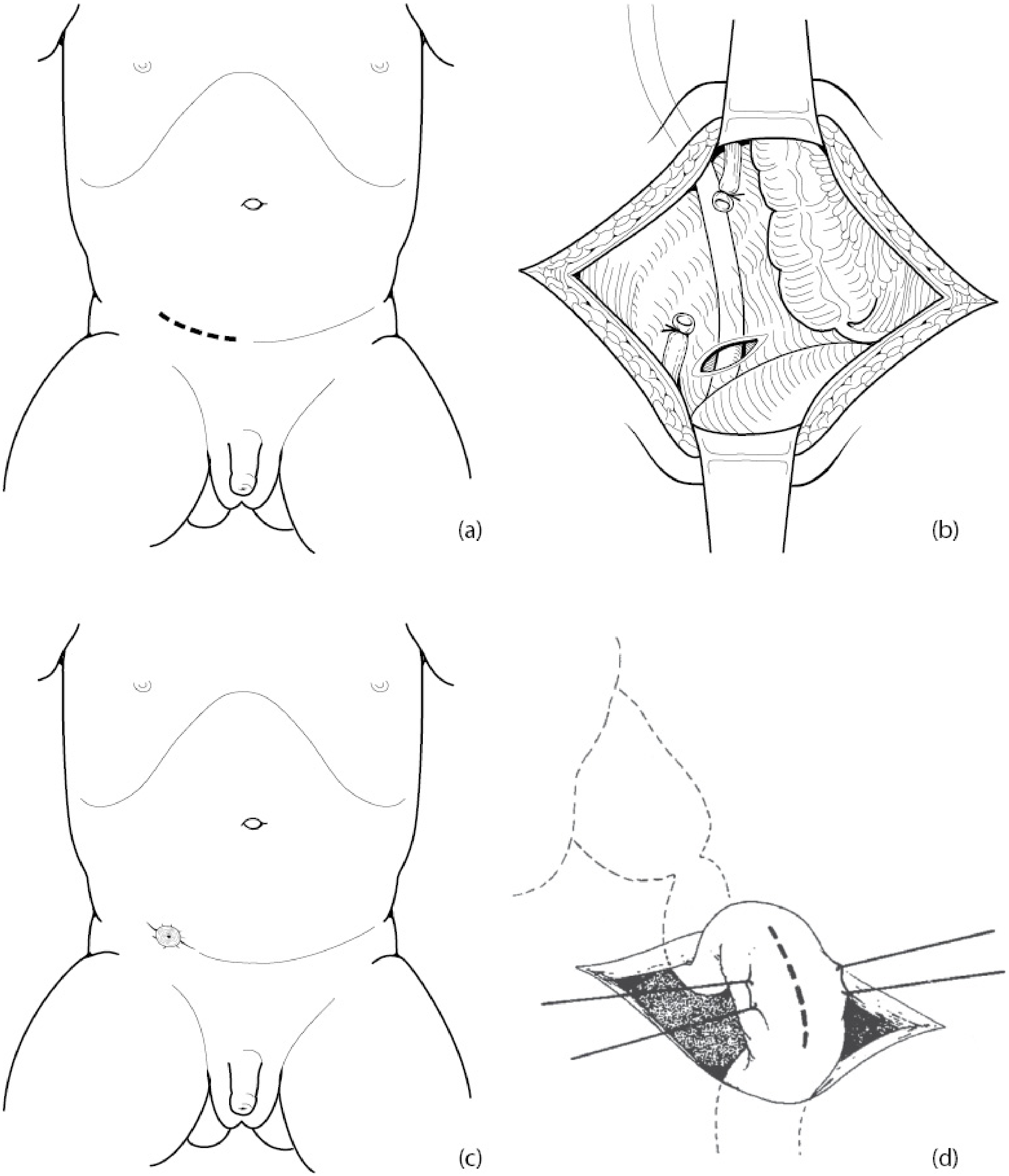
If the vesicostomy is made too low, prolapse of the posterior wall may occur. Stomal stenosis may occur in the long term. A button vesicostomy can have mechanical failure, mucus formation, and stone formation. The button balloon has to be checked every week and the button changed electively every 3–4 months.

Ureterostomy

A cutaneous ureterostomy is used as a temporary measure in carefully selected cases for decompression of the upper tracts. Debate still exists as to its role in PUV with deteriorating upper tracts either as a primary procedure or even after valve fulguration. We have used this method in selected children <1 year of age with solitary kidneys and a vesicoureteric junction obstruction with deteriorating renal function. A ureterostomy may also be performed where malignancy necessitates large pelvic resections. In selected cases of PUV with gross unilateral VUR, a refluxing ureterostomy may be performed on the side of reflux concomitant to valve resection to act as a “pop-off” mechanism and thereby protect the upper tracts. The ureterostomy may be created distally or proximally. The description below is of a distal ureterostomy.

TECHNIQUE

Typically, the child is supine but a more oblique position may be required for a higher diversion. A short inguinal groin crease incision is made on the affected side (**Figure 71.2a**). The site of the incision is modified depending on the site of the intended diversion; however, the approach to the ureter remains extraperitoneal.



71.2a–d

A flap of skin is raised superiorly to gain access to the extraperitoneal space via a muscle split. The peritoneum is reflected medially (**Figure 71.2b**). The ureter is identified by dividing the obliterated umbilical artery.

In cases of end ureterostomy, the ureter is detached close to the bladder and the bladder end is oversewn with 4/0 absorbable suture (e.g. polyglactic acid (Vicryl™) or polydioxanone (PDS™)).

The ureter is brought out of the groin crease incision and the mucocutaneous anastomosis creating a small everted nipple is performed with interrupted 6/0 PDS suture (**Figure 71.2c**).

If a loop ureterostomy is performed, the ureter is opened longitudinally and mucocutaneous anastomosis of both limbs is performed (**Figure 71.2d**).

COMPLICATIONS

Stomal stenosis or stomal prolapse may occur. In cases of dilated tortuous and adynamic upper tracts, good drainage may not be achieved with a ureterostomy and an indwelling catheter or intermittent catheterization of the stoma may be necessary.

Ileal conduit

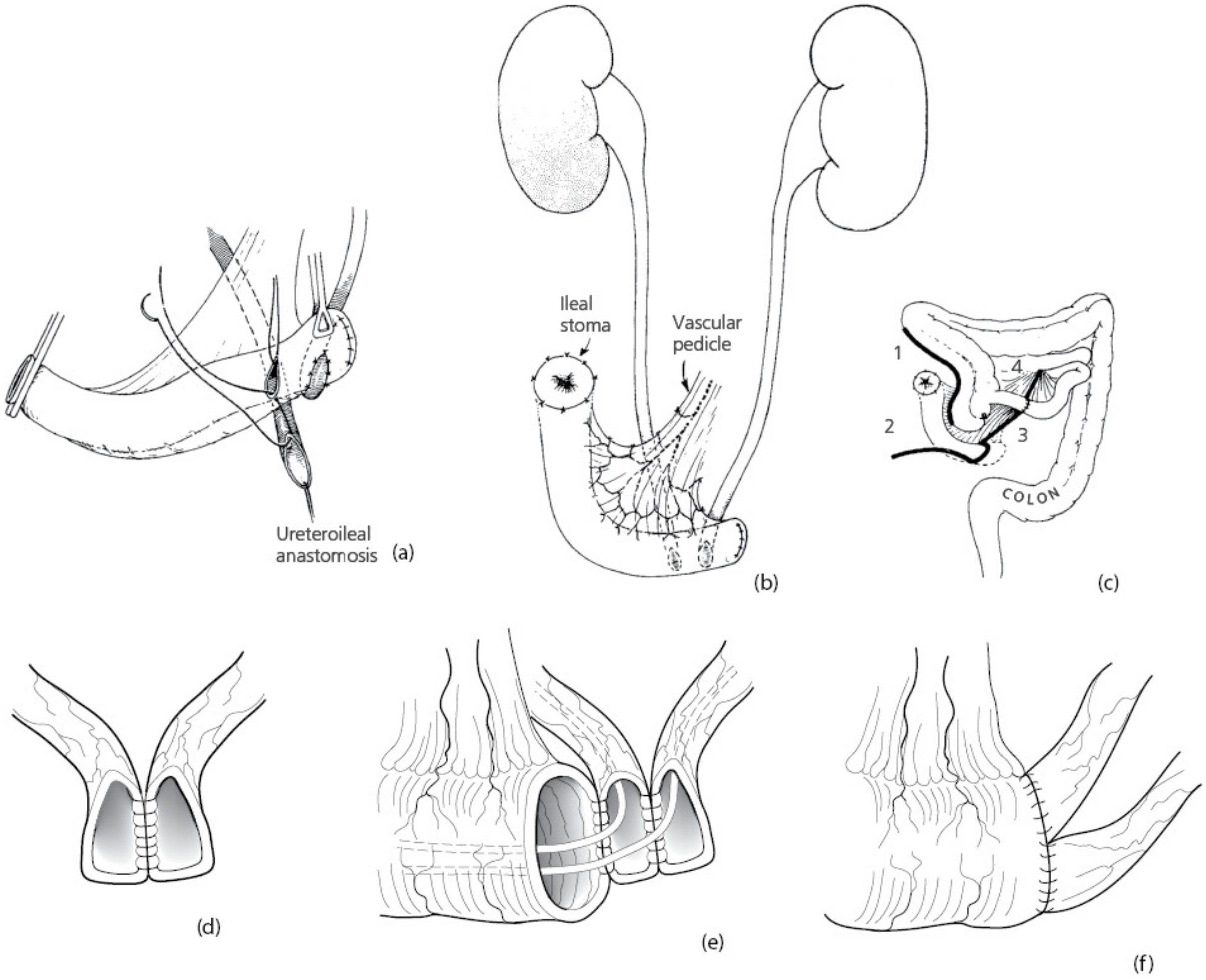
An ileal conduit diversion is very uncommon nowadays in children because of the high incidence of complications. However, in very carefully selected cases, this may be a useful option, for example in children with neuropathic bladder or pelvic malignancy who, along with their carers, may prefer this to major urinary tract reconstruction and continent diversion in the short to medium term.

TECHNIQUE

The ileum can be approached through the same incision as that for the primary procedure if the diversion is being performed concomitantly. A lower midline incision would be used otherwise.

A short, 6–10 cm, segment of ileum on its mesentery is isolated and the proximal end is oversewn with a 4/0 or 5/0 PDS suture.

Following restoration of intestinal continuity, the ureters are spatulated and implanted in the conduit in a submucosal tunnel (**Figure 71.3a**), and the anastomosis is completed with 6/0 PDS interrupted sutures (**Figure 71.3b**). The distal end of the ileal loop is brought out at a suitable premarked position in the right iliac fossa and everted to create a nipple using 5/0 or 6/0 PDS sutures (**Figure 71.3c**). Two 4 Fr or 6 Fr “feeding” tubes are left indwelling within the implanted ureters and exiting via the stoma. If the ureters are dilated, the spatulated ends may be anastomosed to form a common channel (**Figure 71.3d**), which is in turn anastomosed to the proximal end of the conduit (**Figure 71.3e and f**).


Ureteroileal anastomosis
(a)
Ileal stoma
Vascular pedicle
(b)
1
2
3
4
COLON
(c)
(d)
(e)
(f)

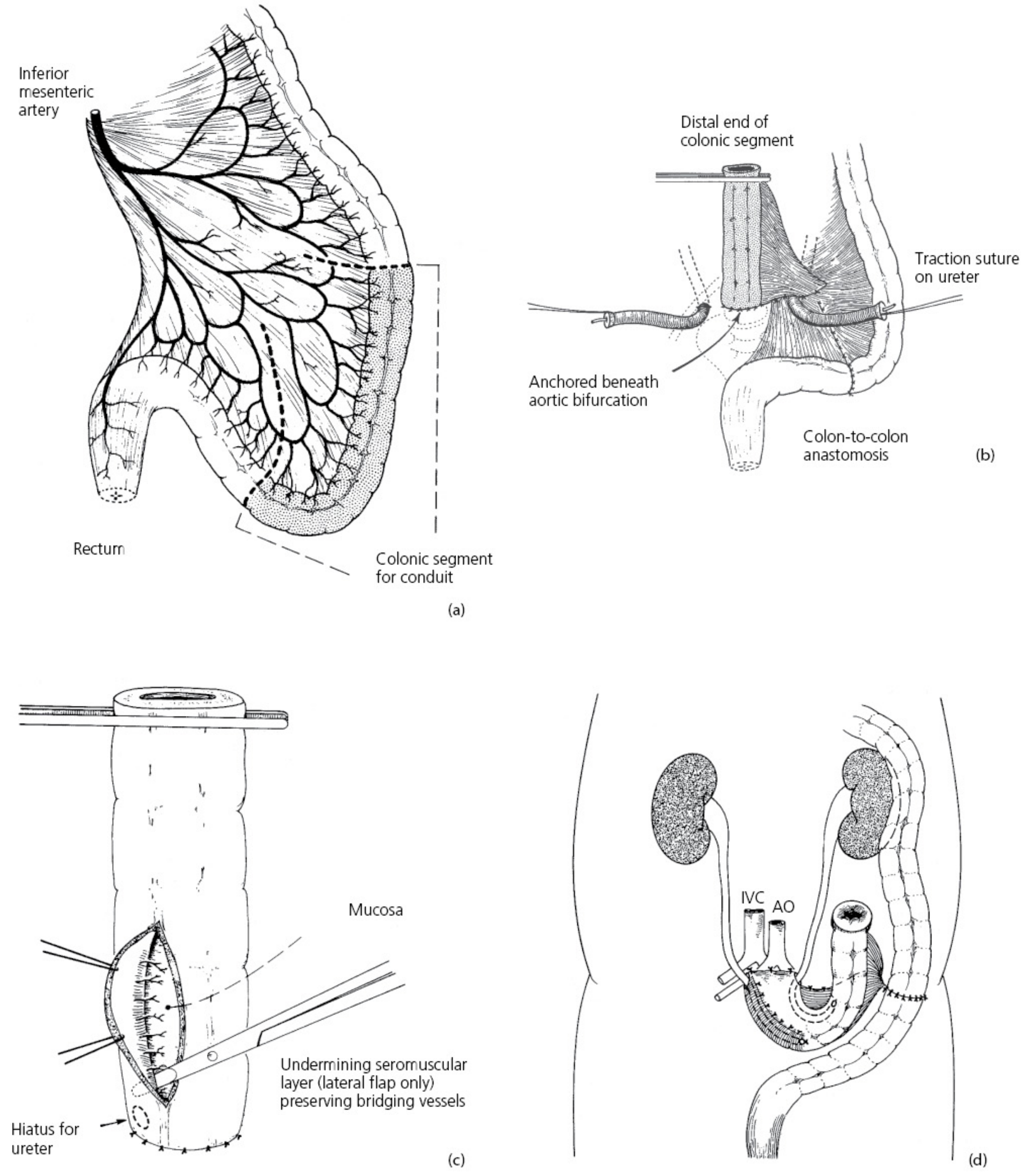
71.3a–f

The ureteric stents may be removed 10–14 days after surgery.

Colonic conduit

TECHNIQUE

A suitable segment of colon, preferably the sigmoid, is isolated on its mesentery and intestinal continuity is restored (**Figure 71.4a and b**). The distal end is oversewn and the ureters are spatulated and implanted in the conduit in a non-refluxing manner (**Figure 71.4c**). The distal end of the conduit is brought out as a stoma at a predetermined, premarked site on the abdominal wall and everted to create a nipple using 5/0 or 6/0 PDS sutures (**Figure 71.4d**).


Inferior mesenteric artery
Rectum
Colonic segment for conduit
(a)
Distal end of colonic segment
Traction suture on ureter
Anchored beneath aortic bifurcation
Colon-to-colon anastomosis
(b)
Mucosa
Undermining seromuscular layer (lateral flap only) preserving bridging vessels
Hiatus for ureter
(c)
IVC
AO
(d)

71.4a–d

CONTINENT URINARY DIVERSIONS

Although most surgery for continent urinary diversion is performed by the open route, there are increasing reports where such interventions are carried out by minimally invasive (laparoscopic or robotic) techniques.

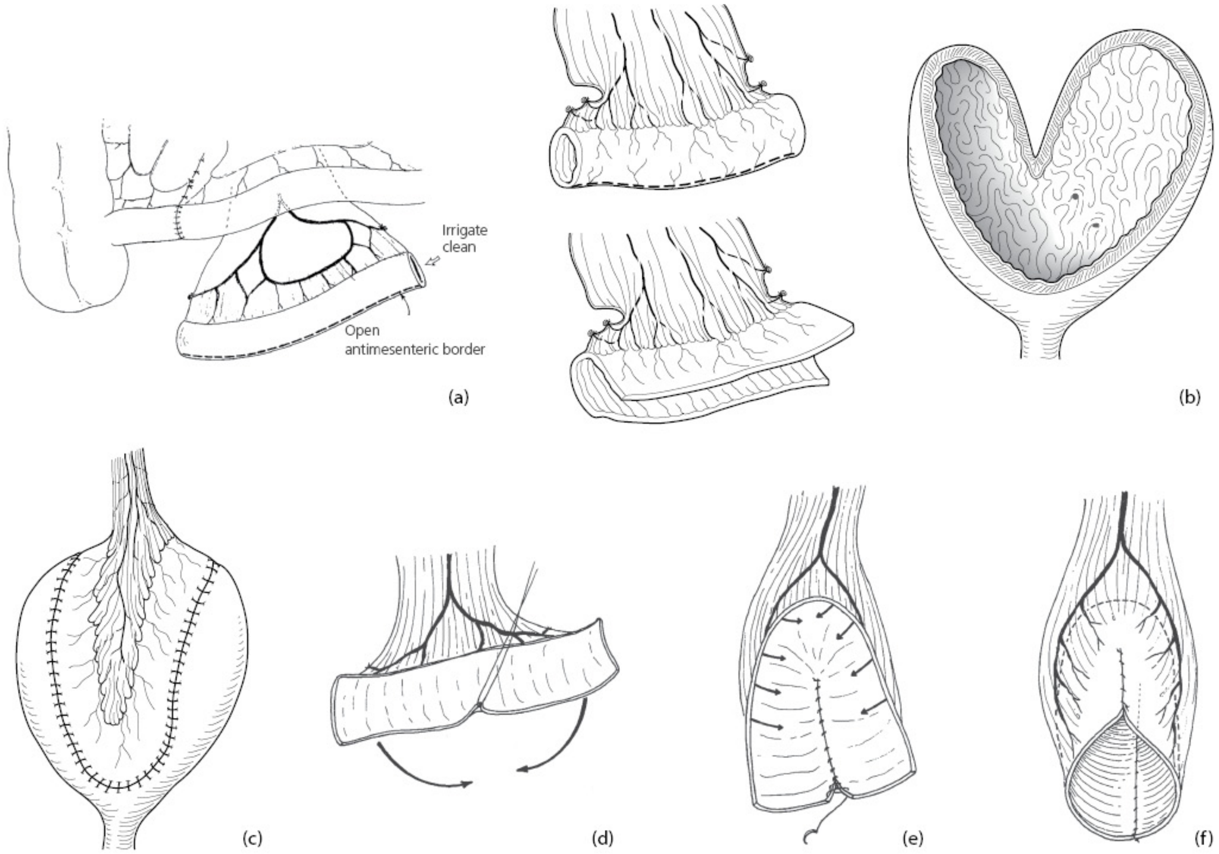
Surgery of the reservoir: Ileocystoplasty

A detubularized segment of ileum is usually used for bladder augmentation. In cases where the mesentery is too short to reach the pelvis (e.g. gross kyphosis) colon may be used. A bladder outlet procedure may be performed simultaneously to increase outlet resistance.

TECHNIQUE

Either a Pfannenstiel or a midline incision can be used.

Initial extraperitoneal mobilization of the bladder is carried out circumferentially down to the region of the bladder neck. A small opening is made in the posterior aspect of the reflected peritoneum and the cecum and terminal ileum are delivered. If an appendicovesicostomy is to be performed, mobilization of the appendix and mesentery can be performed at this stage. A suitable segment of ileum approximately 25 cm long and 20 cm from the ileocecal junction is isolated on its mesentery and intestinal continuity is restored with 5/0 PDS™ sutures (**Figure 71.5a**). The ileum is detubularized along its antimesenteric border. The peritoneum is closed, thereby extraperitonealizing the segment of ileum and bladder. The bladder is opened from one ureteric orifice to the other in either the coronal or sagittal plane (**Figure 71.5b**). The appendix can be reimplanted in the bladder at this stage and a *clam cystoplasty* performed with continuous 3/0 PDS sutures (**Figure 71.5c**). Just before completion of the anastomosis, a large-bore 16 Fr or 18 Fr Foley catheter is inserted suprapubically. If an appendicovesicostomy has been created, an indwelling catheter is left within this conduit. If the bladder is very small, thick-walled, and contracted, it may be excised, leaving only the bladder plate comprising the trigone and the ureteric orifices. In this case, the ileal patch may be reconfigured as a pouch, which can be sutured to the bladder remnant (**Figure 71.5d–f**).



71.5a–f

POSTOPERATIVE CARE

The suprapubic catheter is left on free drainage for 3 weeks with bladder washouts as required. A regime of clamp and release is then instituted prior to commencing clean intermittent catheterization either urethrally or via the Mitrofanoff channel. All patients should have periodic renal ultrasound scans and regular estimation of biochemical parameters, including acid–base status.

COMPLICATIONS

Risks include mucus blockage, urinary infections, bladder calculi, persistence of a high-pressure neoreservoir, metabolic and acid–base disturbances, and the possibility in the long term of the development of adenocarcinoma.

Surgery of the reservoir: Colocystoplasty

The segment used may be detubularized sigmoid colon, the transverse colon, or the ileocecal and ascending colon. The principles of the technique are similar to those described for ileocystoplasty, as are the postoperative care and complications.

The Mitrofanoff principle

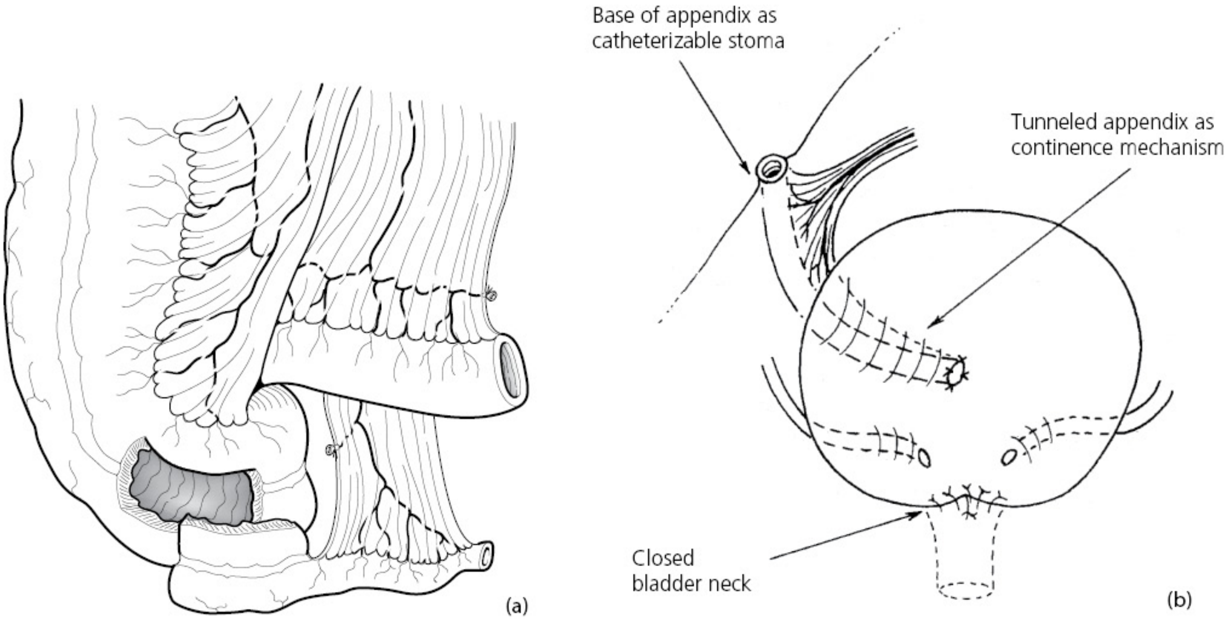
In 1911, Coffey described the flap valve technique of ureteral reimplantation in ureterosigmoidostomy. This principle was first used by Paul Mitrofanoff in 1980 to create a continent, catheterizable appendicovesicostomy. The main components of the Mitrofanoff technique are:

- A narrow, supple conduit brought to the skin as a catheterizable stoma
- An antirefluxing connection between the conduit and the reservoir (flap valve)
- A large, low-pressure urinary reservoir
- An antirefluxing mechanism between the upper urinary tract and the reservoir
- Intermittent catheterization to allow effective, regular, low-pressure emptying of the reservoir

Appendicovesicostomy

TECHNIQUE

A Pfannenstiel or lower midline incision may be used. Firstly, assessment is made of the length of the appendix and its suitability as a conduit. The appendix is mobilized with its mesentery and detached from the cecum, which is then closed. A cuff of cecum may be mobilized with the appendix and tubularized to gain additional length, if necessary (**Figure 71.6a**). The tip of the appendix is incised and a 12 Fr feeding tube is inserted to confirm patency and diameter of the lumen. The lumen is irrigated with dilute Betadine™ solution. The distal end is then tunneled into the bladder in a submucosal tunnel for 3–4 cm to maintain a 5 : 1 ratio and ensure a competent flap-valve mechanism (**Figure 71.6b**). This is then anchored to the muscle and mucosa of the bladder using an absorbable suture. The rest of the circumference of the appendix is sutured to the bladder mucosa using absorbable sutures. The appendix is also anchored at its entry into the bladder.


Base of appendix as catheterizable stoma
Tunneled appendix as continence mechanism
Closed bladder neck
(a)
(b)

71.6a,b

The proximal end of the appendix is brought out as a stoma by the shortest and straightest route possible to enable easy catheterization. The site of the stoma will therefore be variable. The umbilicus may be used to site the stoma in certain children for easy access, or a VQZ stoma technique for abdominal Mitrofanoff stomas can be used. An indwelling 10 Fr or 12 Fr Jacques catheter is left for 3 weeks and is followed by clamp and release and then clean intermittent catheterization.

COMPLICATIONS

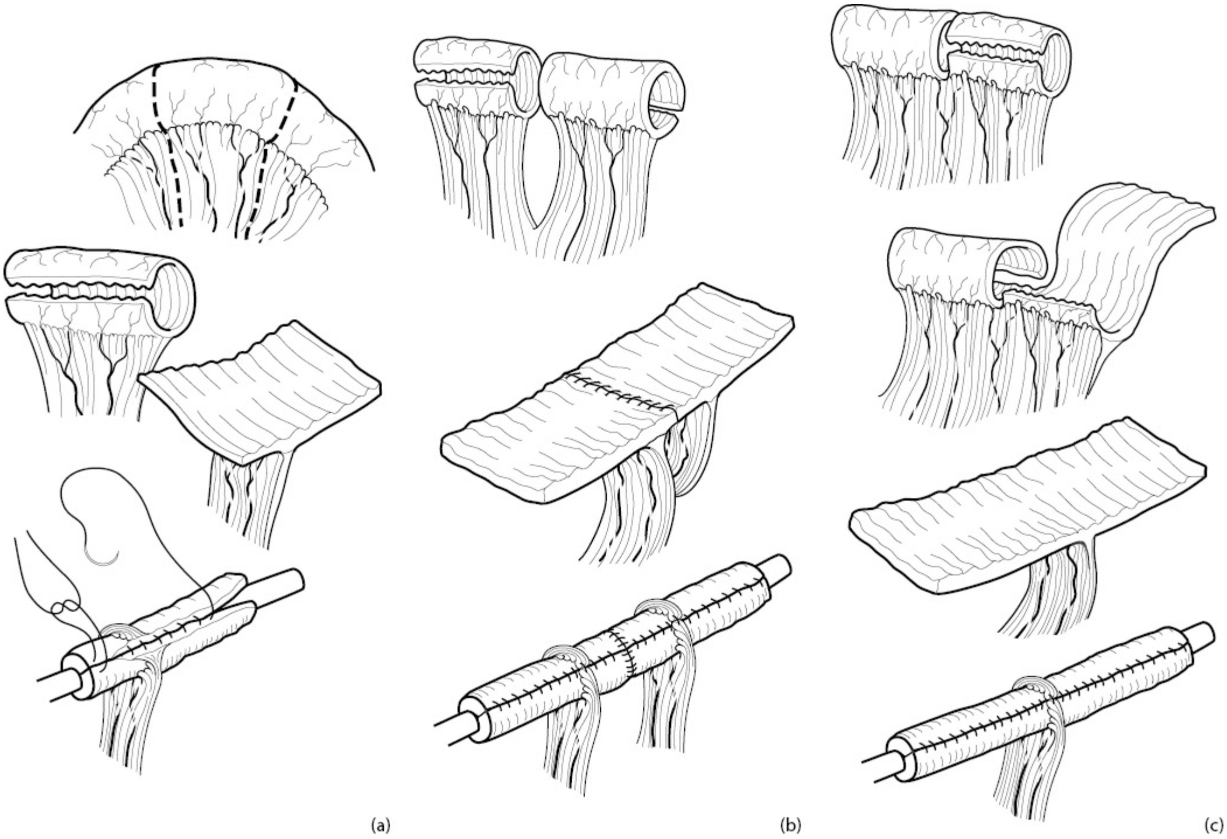
Problems with the conduit include stomal stenosis, difficulty in catheterization leading to the creation of a false passage, and the development of calculi.

Ileovesicostomy

Where the appendix is absent or not suitable, a segment of ileum can be utilized to create a transverse ileal tube. This is the preferred second-line option for creating the Mitrofanoff channel, especially when ileum is used for bladder augmentation.

TECHNIQUE

A segment of ileum approximately 1.5–2 cm in width adjacent to that used for the cystoplasty is isolated on its mesentery and opened longitudinally on its antimesenteric border. The position of the mesentery along the tube can be moved depending on the position of the longitudinal opening. This tube is then tubularized transversely over a 12 Fr catheter (**Figure 71.7a**). In patients with a long distance between the reservoir and the abdominal wall or the umbilicus, two adjacent segments can be isolated, tubularized, and anastomosed (Full Monti, **Figure 71.7b**). A similar technique has been described by Casale using a single piece of bowel to gain additional length (**Figure 71.7c**). The tube is then tunneled into the reservoir as described above to create the Mitrofanoff flap valve.



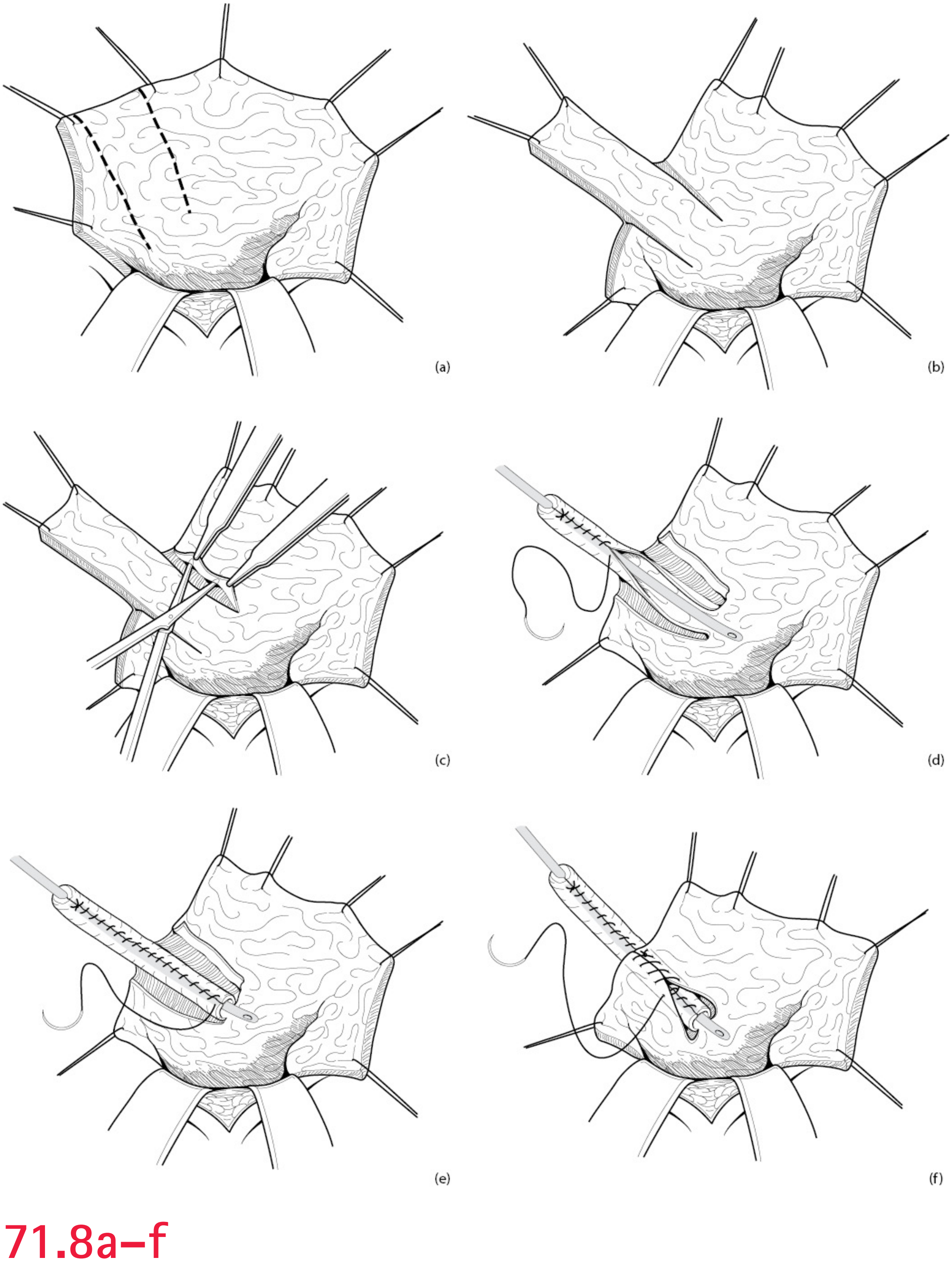
71.7a–c

Detrusor tube vesicostomy

This technique involves a segment of tubularized bladder with an antireflux flap valve constructed primarily of mucosa. Patients must have a large bladder capacity to be considered suitable.

TECHNIQUE

A broad-based rectangular flap (approximately 3 cm × 7 mm) of full-thickness detrusor muscle and mucosa is raised on its vascular supply from the superior vesical pedicle (**Figure 71.8a–c**). This is tubularized in two layers (mucosa and muscle) over a 12 Fr catheter. The proximal mucosal end is further tubularized within the bladder for about 2 cm and covered by laterally raised adjacent mucosal flaps (**Figure 71.8d–f**).



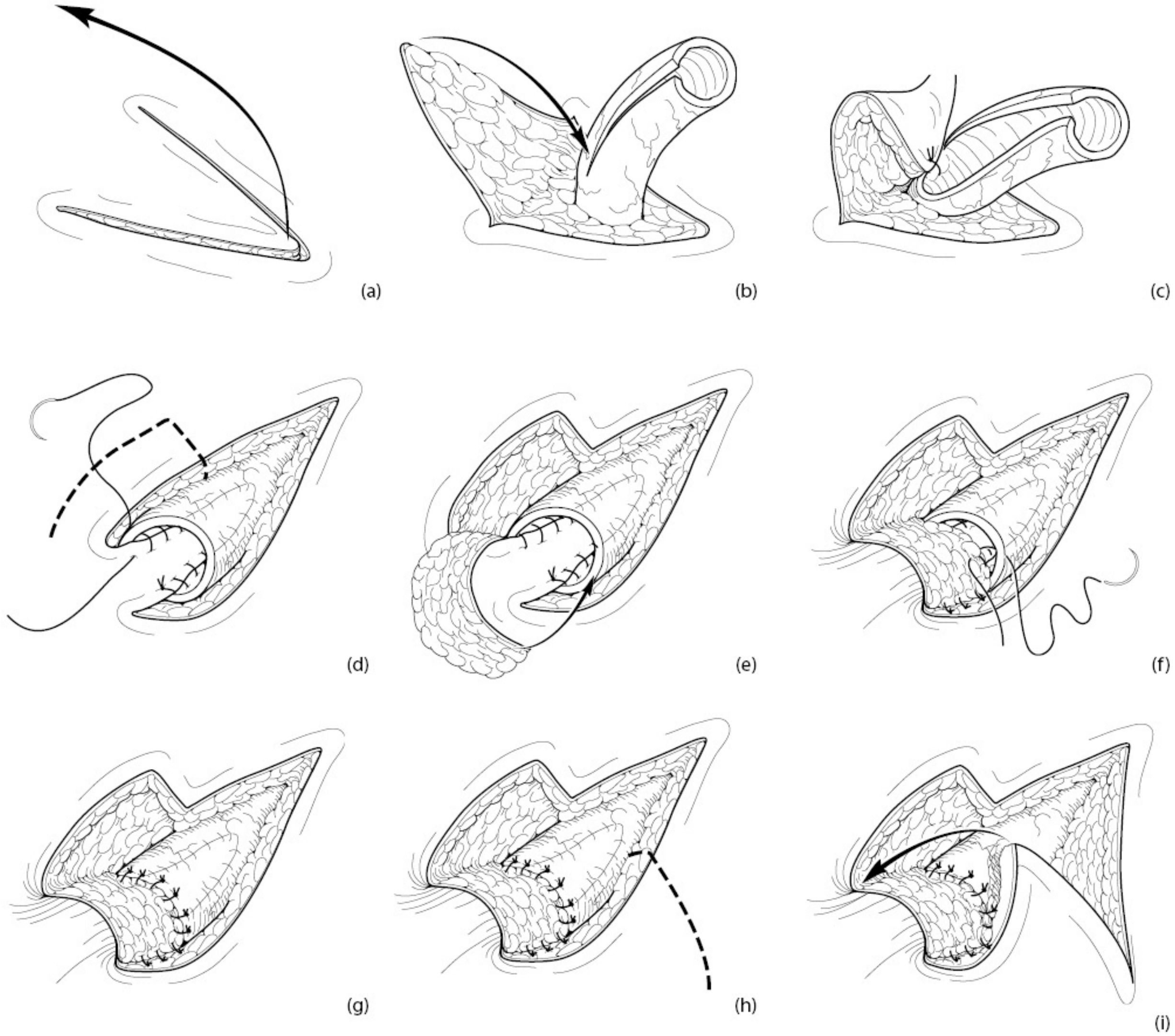
71.8a–f

The VQZ stoma

The VQZ plasty for the cutaneous opening into a Mitrofanoff conduit has several advantages over the standard V flap or flush stoma in that it is discrete and there is no exposed mucosa and hence less likelihood of contact bleeding.

TECHNIQUE

A V flap is raised with its base in the region of the exiting appendix (**Figure 71.9a**). A generous opening is made into the layers of the anterior abdominal wall and the appendix is delivered. The abdominal incision is then closed. The appendix is incised along its antimesenteric border and the flap is sutured in place with 5/0 PDS along its entire length up to its angle on its superior border, but stopping 5–10 mm short on its inferior border (**Figure 71.9b–d**). The Q flap is then raised and rotated, and sutured into place along the rest of the appendicular margin and the remaining inferior border of the V flap (**Figure 71.9e–g**). The mucosa is therefore entirely covered. A standard Z plasty is performed to rotate the adjacent skin to cover the defect (**Figure 71.9h and i**). Subcutaneous 3/0 PDS sutures may be required at the angles of the Z plasty, which will be under some tension. The rest of the skin is closed with 5/0 or 6/0 PDS sutures.



71.9a–i

FURTHER READING

Adams MC, Rink RC. Augmentation cystoplasty. In: Gearhart J, Mouriquand P, Rink R (eds). *Pediatric Urology*. Philadelphia, PA: WB Saunders; 2010: pp. 748–61.

Farrugia MK, Malone PS. Educational article: The Mitrofanoff procedure. *J Pediatr Urol* 2010; 6: 330–7.

Leslie JA, Dussinger AM, Meldrum KK. Creation of continence mechanisms (Mitrofanoff) without appendix: The Monti and spiral Monti procedures. *Urol Oncol* 2007; 25: 148–53.

Metcalfe PD, Cain MP. Incontinent and continent urinary diversion. In: Gearhart J, Mouriquand P, Rink R (eds). *Pediatric Urology*. Philadelphia, PA: WB Saunders; 2010: pp. 737–48.

Palmer BW, Kropp BP. Augmentation cystoplasty and diversion. In: Wilcox DT, Godbole PP, Koyle MA (eds). *Pediatric Urology, Surgical Complications and Management*, 2nd ed. London: Wiley-Blackwell;

2015: pp. 377–84.

Traxel EJ, Minevich EA, Noh PH. A review: The application of minimally invasive surgery to pediatric urology: Lower urinary tract reconstructive procedures. *Urology* 2010; 76:115–20.

72

Renal calculi

ELENI PAPAGEORGIOU AND NAIMA SMEULDERS

INTRODUCTION

The incidence of urolithiasis in children is increasing. Ultrasound (US) remains the mainstay of imaging. As a result of the dramatic reduction in radiation of ultra-low-dose non-contrast computed tomography (CT) using the newer generation scanners and focused on the area of interest alone, CT is increasing in popularity, particularly for complex stones. A metabolic cause for stones is found in a significant proportion and appropriate investigation is therefore essential for all children with urolithiasis. However, these tests should not delay surgery to clear stones. Sterilizing urine prior to intervention is essential to reduce the risk of overwhelming postoperative sepsis.

Minimally invasive surgical techniques have replaced open surgery for stone clearance.

EPIDEMIOLOGY

Stone formation in the urinary tract affects approximately 5–10% of the population. Children account for just 1–3% of all stone patients. The incidence of urolithiasis shows marked geographical variation: While it is very rare in Greenland and Japan, a “stone belt” extends from the Balkans across Turkey, Pakistan, and north India.

Overall, boys are affected twice as often as girls and tend to present at a younger age. The average age at presentation is 4.4 years for boys and 7.25 years for girls. Presenting features are macroscopic hematuria, urinary tract infection (UTI), and/or abdominal pain; some children (~13%) appear to be asymptomatic (**Table 72.1**). Obstruction related to calculi may result in pyonephrosis, perinephric abscess, or progressive pyelonephritis and renal loss.

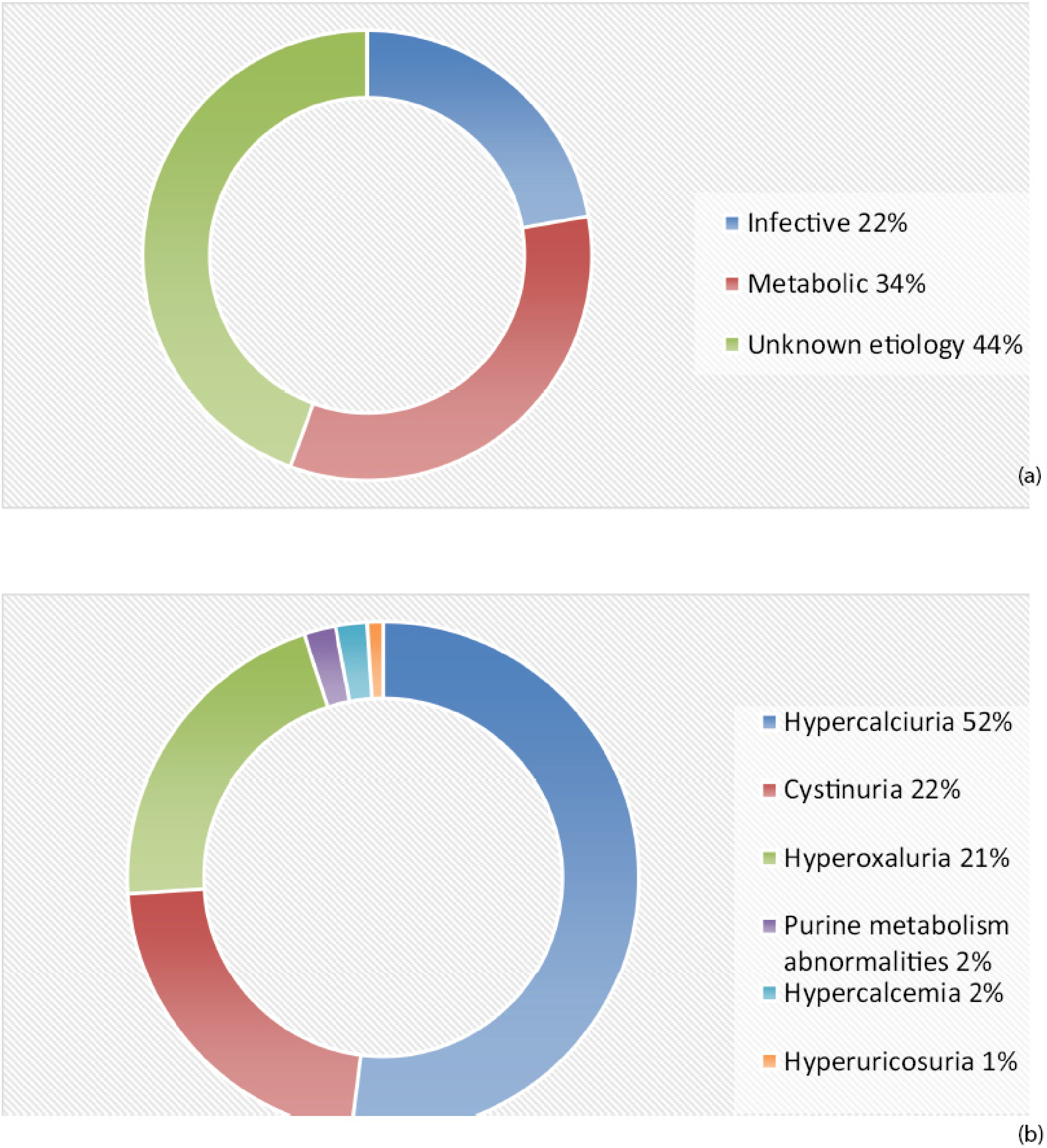
Table 72.1 **Presenting features of urolithiasis**

Presenting features	**Incidence (%)**
UTI	36
Abdominal pain	32
Macroscopic hematuria	27
Painful hematuria	13
Asymptomatic	13
Coincidental finding	11
Screening	1.8
Acute kidney injury	1.4

Source: Issler et al. *BMC Nephrol* 2017; 18: 136.

ETIOLOGY

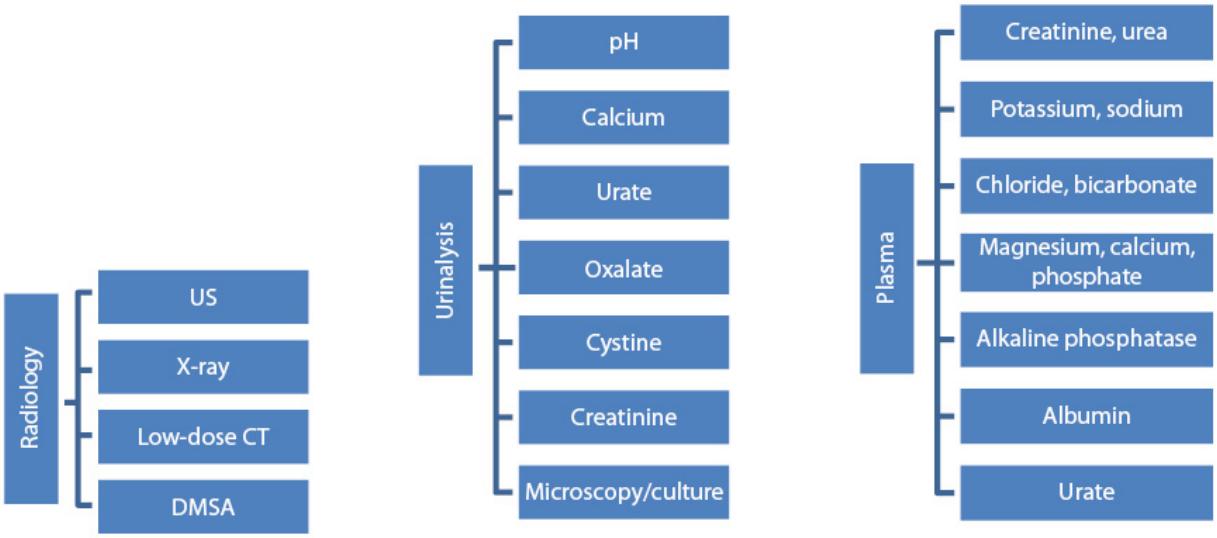
Until recently, most renal calculi in children were considered infective in origin. The organisms most commonly associated with infective calculi are the urea-splitting *Proteus* spp. and *Escherichia coli*. Infective calculi are usually soft, contain organic matrix, and opacify poorly. They usually consist of calcium, magnesium, and ammonium phosphate. Today, the epidemiology appears to have changed. In a review from our institution (1993–2015) a metabolic abnormality was detected in 34% of children with urolithiasis; 22% of stones were classified as infective, while in 44% of patients no etiological factor could be identified (**Figure 72.1a**). Hypercalciuria (52%) was the most common metabolic abnormality, followed by cystinuria (22%), hyperoxaluria (21%), unclassified hypercalcemia (2%), abnormalities in the purine metabolism (autosomal recessive adenine phosphoribosyl transferase deficiency, 2%), and hyperuricosuria (1%) (**Figure 72.1b**).



72.1a,b (a) Etiology and (b) metabolic etiology of urolithiasis. (After Issler et al. *BMC Nephrol* 2017; 18: 136.)

PREOPERATIVE INVESTIGATIONS AND PREPARATION

See **Figure 72.2**.


Radiology
US
X-ray
Low-dose CT
DMSA
Urinalysis
pH
Calcium
Urate
Oxalate
Cystine
Creatinine
Microscopy/culture
Plasma
Creatinine, urea
Potassium, sodium
Chloride, bicarbonate
Magnesium, calcium, phosphate
Alkaline phosphatase
Albumin
Urate

72.2 Radiological, urine, and blood tests for urolithiasis.

Radiology

The main imaging modality is US, which is able to detect stones throughout the urinary tract, although stones in the mid- or distal ureter may be obscured by bowel gas, especially if the bladder is empty. US should give an assessment of the size, shape, number, and location of calculi. Dilatation proximal to an obstructing stone will also be shown and the presence of debris will raise the suspicion of infection and the need to relieve the obstruction urgently.

Plain abdominal X-ray of the whole urinary tract may be a useful adjunct, but bowel gas may make interpretation difficult in children. Uric acid and cystine calculi are mildly radio-opaque while xanthine and dihydroxyadenine stones are radiolucent.

Nowadays, CT is increasingly indicated in children as the newer scanners have significantly reduced their radiation burden. Furthermore, combining functional imaging, principally dimercaptosuccinic acid (DMSA) scan, to detect renal scarring with SPECT and a low-dose limited CT can guide treatment decisions for those with difficult anatomy and complex or recurrent renal calculi.

Intravenous urogram and CT urogram are rarely needed.

Urinalysis

Microscopy and urine culture are performed. Stones are analyzed for chemical composition. Urine is sent for determination of pH, calcium, urate, oxalate, cystine, and creatinine. These investigations may be performed before surgery or much later when the child is stone-free.

Blood investigations

Creatinine, urea, potassium, sodium, chloride, bicarbonate, magnesium, calcium, phosphate, alkaline phosphatase, albumin, and urate tests are performed. The findings of nephrocalcinosis, bilateral calculi, or recurrent calculi suggest a metabolic abnormality. Nephrocalcinosis may be associated with renal tubular acidosis, hyperoxaluria, and hypercalcemia.

Preparation

In order to reduce the risk of sepsis, urinary infection must be sought preoperatively and treated, as well as appropriate IV antibiotics administered on induction of anesthesia in all.

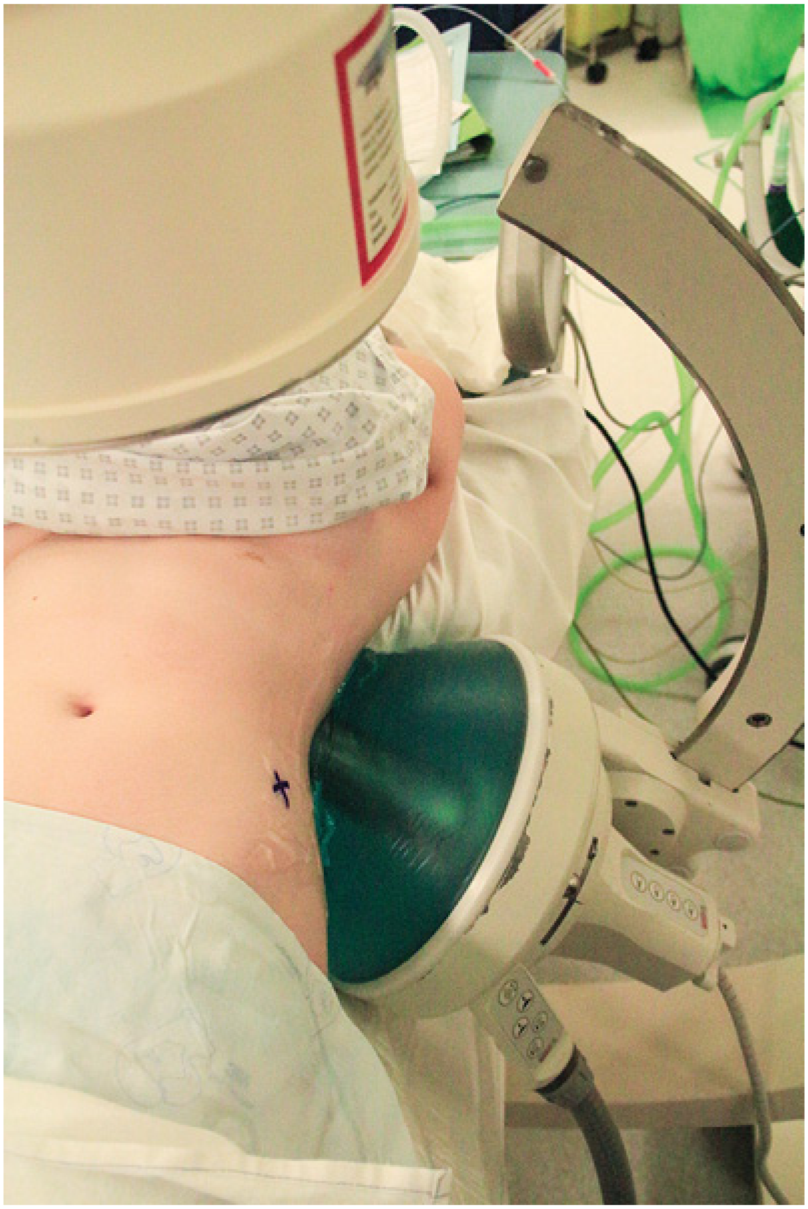
OPERATIONS

As a result of the dramatic advances in technology over the last decades, the less invasive techniques of extracorporeal shock-wave lithotripsy (ESWL), percutaneous nephrolithotomy (PCNL), ureterorenoscopy (URS)/retrograde intrarenal surgery (RIRS) and percutaneous cystolithotomy (PCCL) have superseded open surgery for renal calculi in children. Laparoscopic operation may also be considered in some cases.

The aim of treatment is to render the child stone-free with preservation of maximal renal function and minimal complications. The treatment modality/modalities chosen will depend on the stone burden and etiology, pelvicalyceal location and anatomy, the symptoms and complications caused by the stone(s), as well as comorbidities. All residual fragments are significant, particularly in the presence of infection, but also with an underlying metabolic abnormality. One-third of small residual fragments will regrow over a period of 1–4 years.

Extracorporeal shock-wave lithotripsy

First performed in 1980, the principle of ESWL is to shatter a stone into smaller fragments, which can pass spontaneously through the urinary tract. Pulses of externally generated shock waves are focused by an external parabolic dish onto the stone (**Figure 72.3**). The shock wave was originally generated by an electric spark, but other methods of inducing shock waves including rapid vibration of piezoelectric crystals or electromagnetic diaphragms have since evolved. The newer second-generation machines employ a water cushion rather than a large water bath for coupling (transmission of the shockwaves through the skin), and are combined with more accurate US- and fluoroscopy-guided targeting systems in the third-generation lithotripters. These improvements have resulted in less discomfort, and children may be treated under simple analgesia. Nevertheless, in practice, it is difficult to maintain a child in one position during treatment (typically 30–45 minutes), and general anesthesia or heavy sedation continues to be used in young children and infants.



72.3 ESWL: The patient rests on a water-based cushion. The US probe for concurrent imaging of the targeted stone can be seen within the cushion. Gel facilitates transmission of the shock waves into the body.

In children, stones clear from a lower-pole location as well as from any other renal location, which is in contrast to adults. It is the treatment of choice for single renal pelvic stones up to 20 mm, lower-pole stones up to 10 mm, and proximal ureteric stones.

Success rates are in the order of 80%, although repeat procedures might be needed. In our experience, a stone that has failed to clear after two session of ESWL does not benefit from a third sitting and a different strategy should be explored.

Stone clearance rates are dependent on stone size and composition. Where a CT scan has been performed, Hounsfield units (HU) of density (named after Godfrey Hounsfield (1919–2004), the British Nobel prize-winner engineer responsible for development of CT) have been shown to be an independent predictive factor of clearance: Stones <600 HU have a success rate of 80% compared to 20% for stones of >600 HU. Cystine stones, calcium oxalate monohydrate, and calcium phosphate dihydrate show poorer response to ESWL. Stone clearance is also significantly affected by stone burden, number of stone per renal unit, and a composition of calcium phosphate >70%, but it is not impacted by age, stone location, uni- or bilateral stones, or the patient's ambulatory status.

Side effects such as transient hematuria, renal colic, and petechial hemorrhages are common. Complications of hematoma, sepsis, and stone fragment ureteric obstruction (*Steinstrasse* – the German word meaning "stone street") occur in about 5%.

A recent study, with follow-up extending to over 10 years, has negated previous concerns regarding its impact on immature organs, showing no effect on renal growth or on the incidence of hypertension and diabetes.

Percutaneous nephrolithotomy

PCNL involves needle puncture of the kidney under US or X-ray guidance and dilatation of the tract to allow direct visualization, fragmentation, and removal of renal stones.

It is used to disintegrate and remove:

- Multiple, large, or staghorn calculi
- Calculi in patients with gross spinal deformities where focusing the shock waves onto the stone is difficult
- Moderate-sized calculi without disintegration and risk of residual fragments (e.g. above a long, narrow infundibular neck, in a calyceal diverticulum, or in a horseshoe kidney)

Where residual stones persist after PCNL, these can be cleared subsequently by ESWL, second-look PCNL and/or URS.

Since 1980, different PCNL systems have been developed (**Table 72.2**).

Table 72.2 Development of PCNL systems

PCNL	Year	Tract size (Fr)
Standard PCNL	1980	24–30
Mini PCNL	1997	<20
Micro PCNL	2010	4.8
Ultra-mini PCNL	2012	11–16
Minimally invasive PCNL (MIP)		9.5–30
Super-mini PCNL (SMP)	2012	10–14 with active suction

Bigger tract sizes allow nephroscopes with larger working channels and more efficient lithotripsy tools, as well as larger stone fragments to be lifted out of the kidney. The Amplatz sheath should be ≥3 Fr larger than the nephroscope to permit free backflow of irrigation fluid. Keeping the operative time to less than 100 minutes reduces the risk of life-threatening urosepsis. On DMSA scanning no difference in scarring has been observed between 11 Fr and 30 Fr tracts. The main benefit of decreasing tract size is on hematuria and time to recovery/discharge.

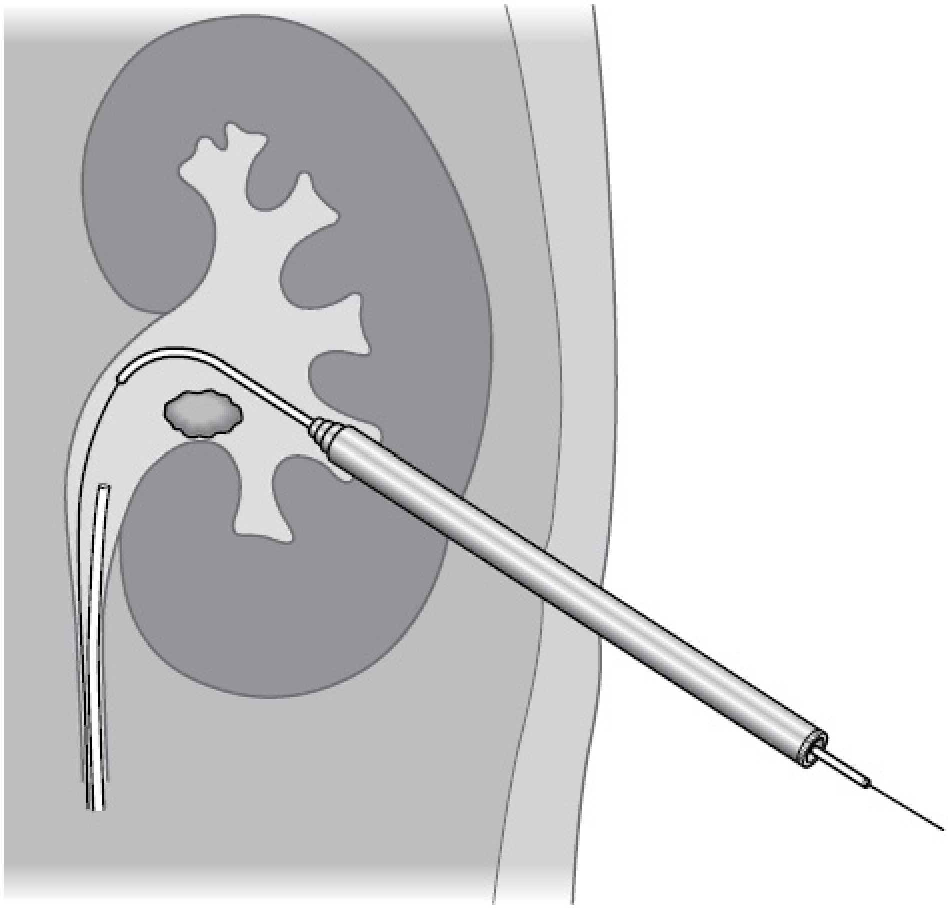
The super-mini PCNL (SMP), recently developed in China, consists of a 7 Fr nephroscope with a 3.3 Fr working channel and a 10, 12 or 14 Fr Y-shaped sheath with integrated suction. It can be used for stones up to 2.5 cm in size, but is especially practical for stones unsuitable for ESWL or those with difficult anatomy. SMP can be used alone, replacing RIRS, or in combination with standard PCNL. Lithotripsy is performed by Holmium YAG Laser and the stone fragments are cleared by powerful built-in suction. Alternatively, a disposable Y-shaped sheath can be used with a ureteroscope.

The PCNL risks are hemorrhage (10%), fever (20%), infection (5%), TUR syndrome, urinoma, hydrothorax/pleural injury, and pelvicalyceal scarring/stricture (0.2%). Hypothermia is a significant risk and all facilities to prevent this should be employed, including warming of the operating room, warming of IV and of irrigation fluid, forced-air warming systems, and the careful application of plastic sticky drapes to prevent the pooling of irrigation fluid around the child.

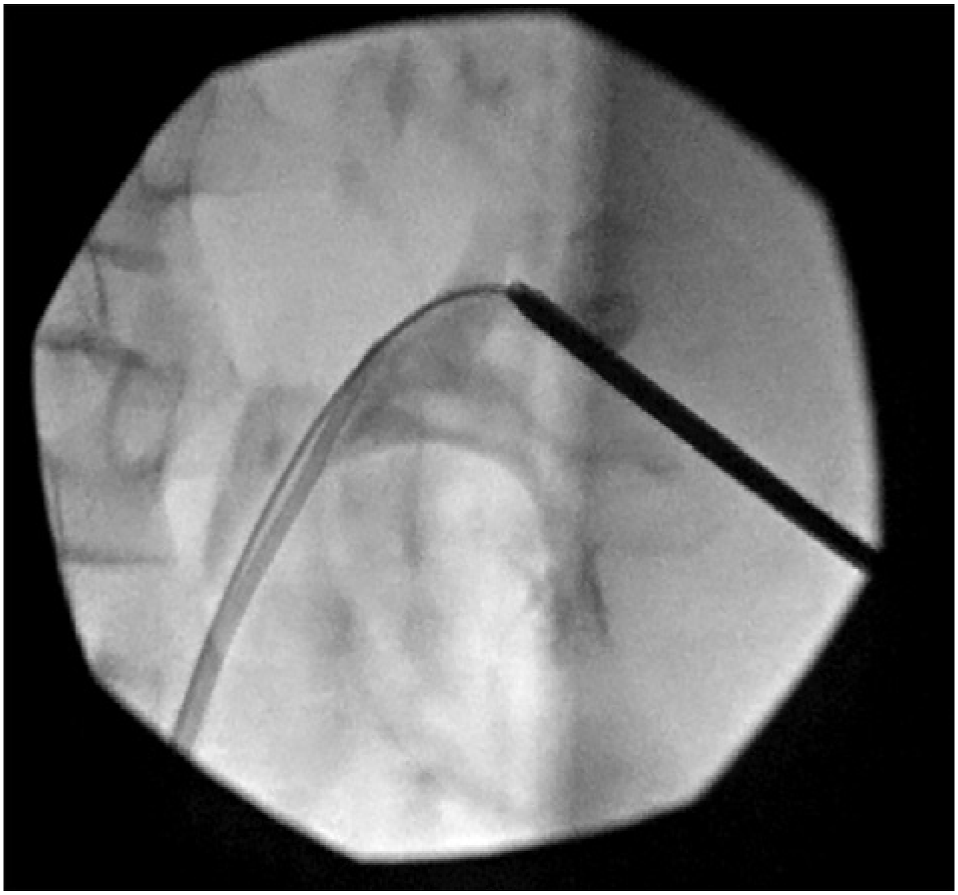
For the procedure, the child is fully anesthetized. In order for the head to be turned to the same side as the kidney to be operated on in the prone position, the anesthetist should be asked to secure the endotracheal tube to the side of the affected kidney.

By cystoscopy, a guidewire is passed retrograde into the ipsilateral ureter. A ureteric catheter is screened to just below the ureteropelvic junction and the guidewire removed. The ureteric catheter is secured to a urethral catheter (e.g. balloon catheter), and both are taped to the leg of the child. The purposes of the ureteric catheter are, first, to enable contrast to be instilled into the pelvicalyceal system and, second, for stone fragments that threaten to migrate down the ureter during the PCNL to be flushed back into the pelvis for extraction. The child is then turned prone and supported by gel pads.

Under US and/or fluoroscopy guidance, an appropriate calyx is punctured and an angled hydrophilic guidewire advanced down the ureter (**Figures 72.4 and 72.5**). After exchanging the wire for a stiffer guidewire, the needle is removed and the tract dilated. Different systems for dilation exist, but the authors prefer the telescopic metal dilatation system.

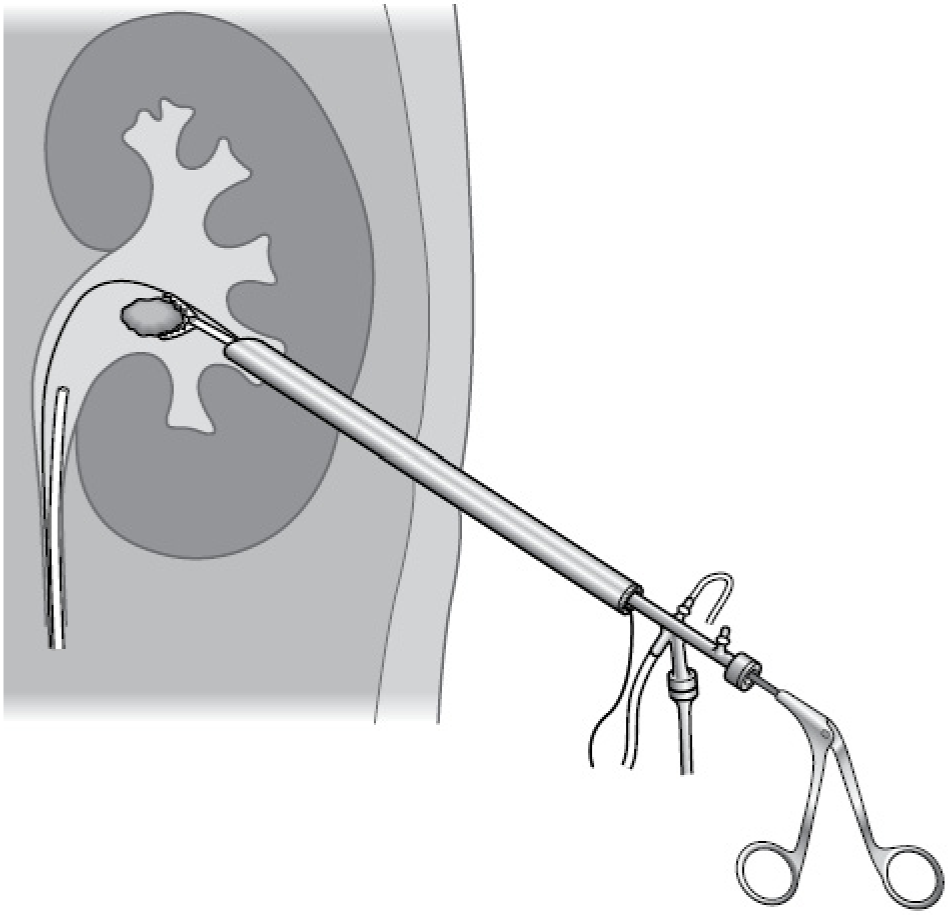


72.4

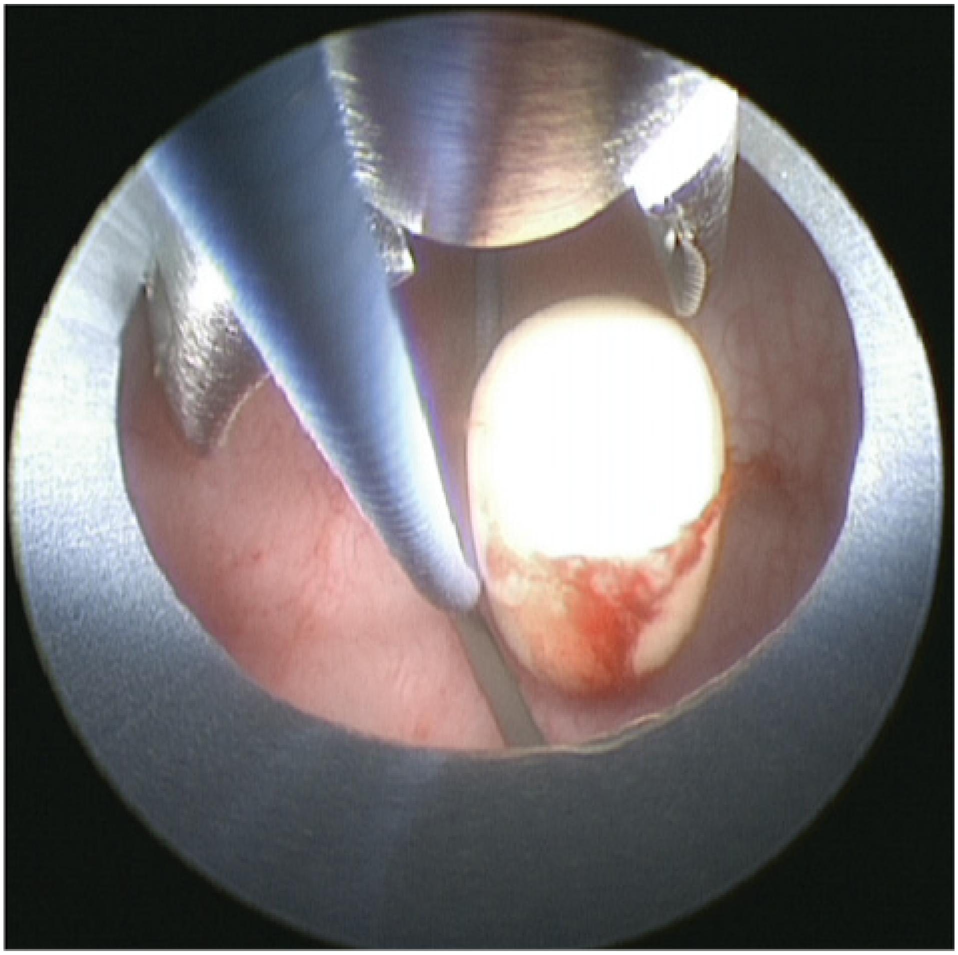


72.5 PCNL: An interpolar calyx has been accessed and dilated under fluoroscopic guidance over a guidewire, which has been passed alongside the retrograde catheter down the ureter.

An Amplatz sheath is placed over the dilator. The size of the sheath chosen will depend on the size of the child, the locations and volume of the stones, and the telescope to be used. The chosen tract size must permit free backflow of irrigation fluid around the nephroscope (e.g. 24 Fr Amplatz sheath for 20.8 Fr nephroscope) to prevent hydrostatic pressure damage to the kidney. By nephroscopy, a small stone can be removed through the sheath with graspers (**Figures 72.6 and 72.7**). The assistant should secure the Amplatz sheath during extraction.

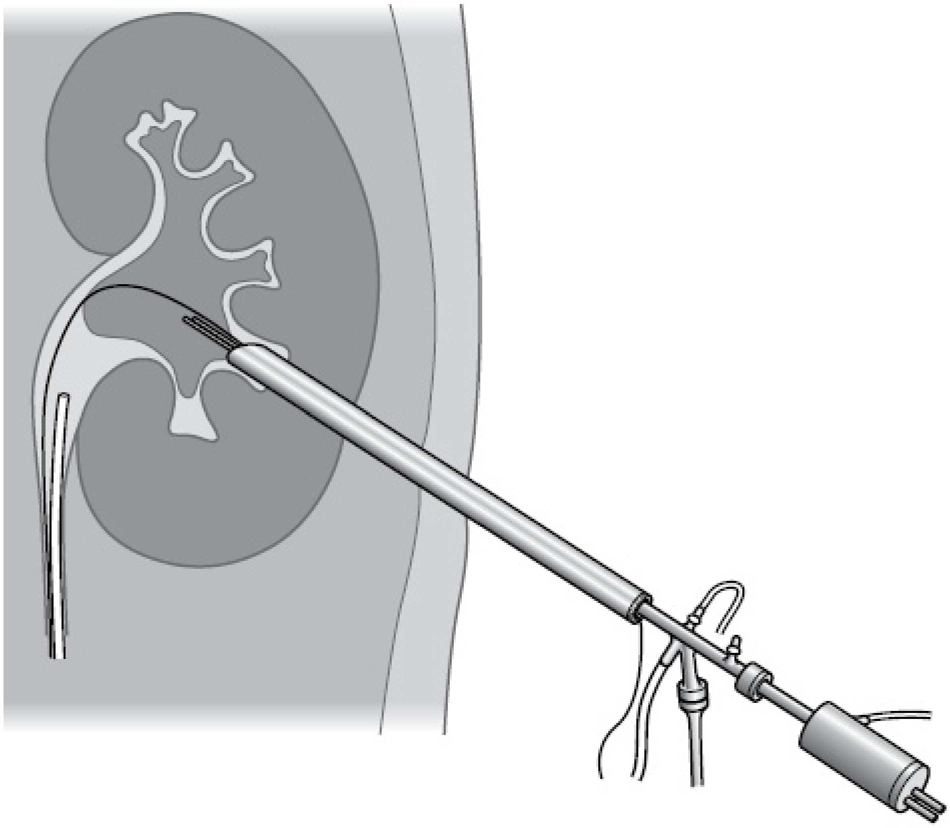


72.6

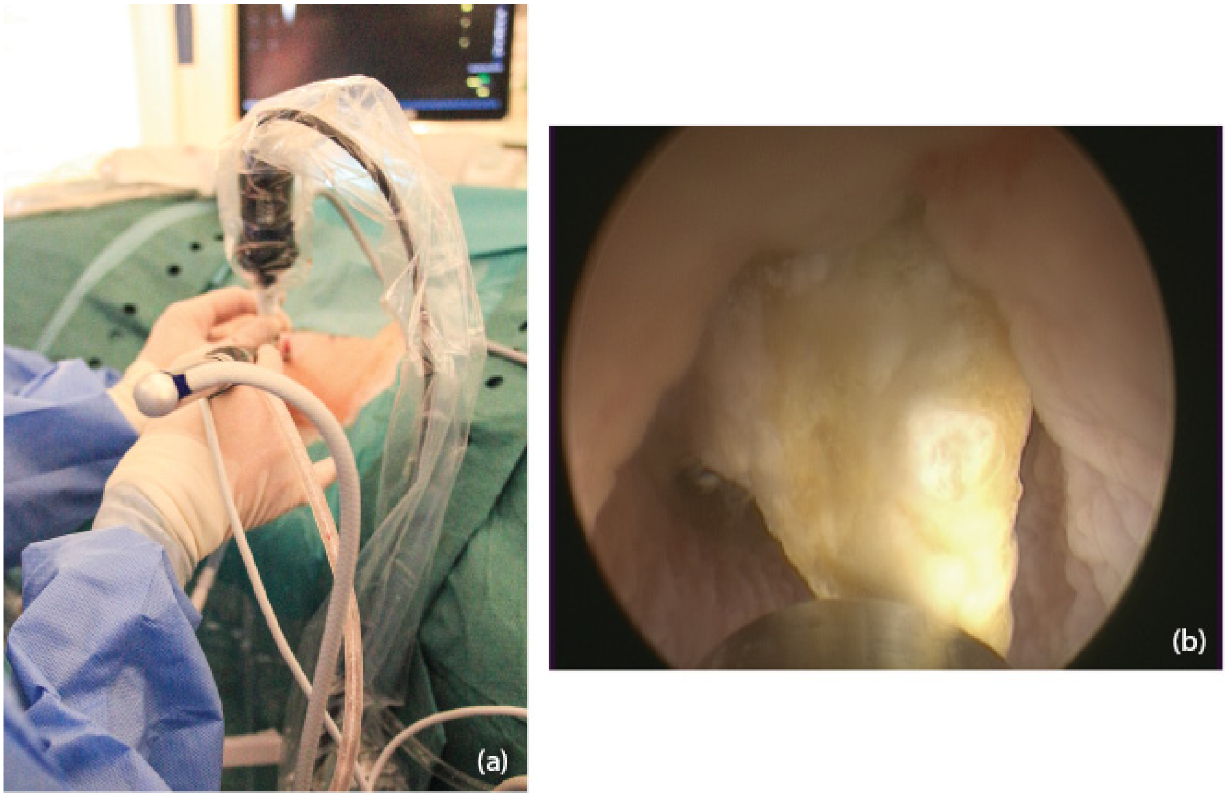


72.7 PCNL: Grasping forceps are used to lift small stones or stone fragments out of the kidney through the Amplatz sheath.

A staghorn calculus can be disintegrated under direct vision using an US and/or lithoclast probe (**Figures 72.8 and 72.9**). For the most expedient fragmentation, US with ballistic system combined with continuous suction, requires a 10 Fr working channel. For complex stones, multiple punctures into different calyces may be required, although not all will need dilatation. Stones may be "snookered" out of the calyx into the renal pelvis using the Kellett needle under US visualization with or without a wire. Where this fails, dilating to a 10–14 Fr enables a concurrent SMP. Finally, flexible operating telescopes may allow access to further stones for fragmentation by laser, or, using a basket, repositioning in line with the primary tract for prompt extraction.



72.8



72.9a,b PCNL: For stone fragmentation, the lithoclast is passed through the working channel of the nephroscope (a) onto the stone (b).

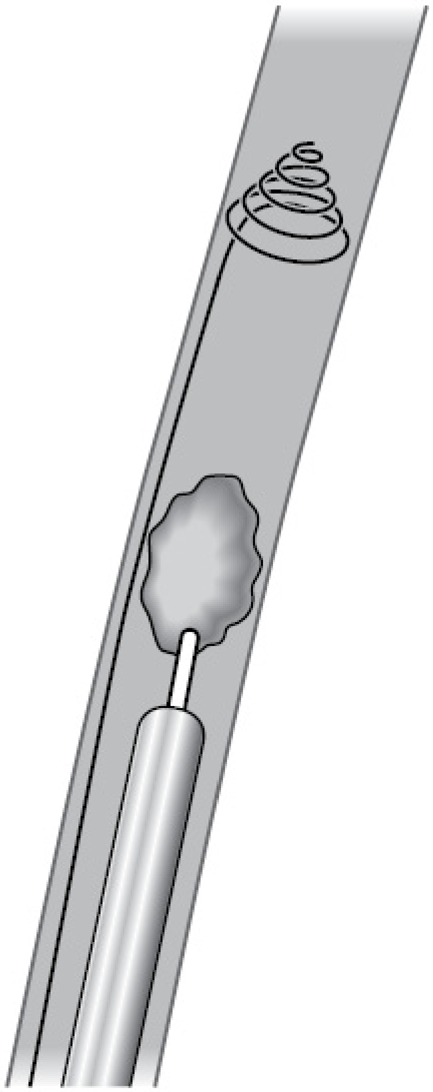
POSTOPERATIVE CARE

Nephrostomy drainage is advised for all except simple “lift-out” PCNLs for 24–48 hours. The average hospital stay is 4 days. Stone-free rates are 90–100%. SM can be tubeless if there is no bleeding.

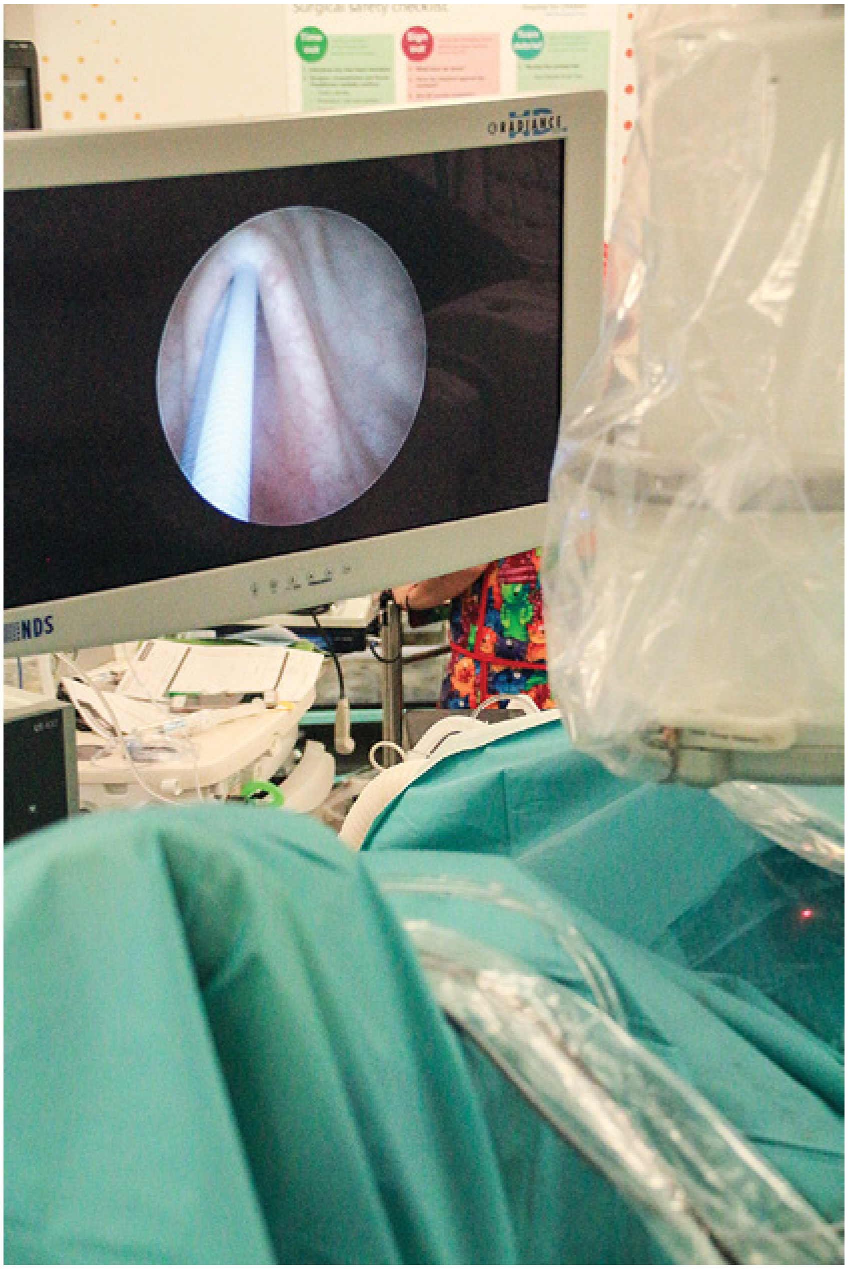
Ureterorenoscopy

Small (4.5–8.5 Fr) rigid or flexible telescopes can be passed through the bladder into the ureter and kidney, enabling fragmentation and extraction of ureteric and small renal stones. By cystoscopy, a guidewire is inserted into the ureter and placed into the renal pelvis under fluoroscopy guidance. A retrograde ureteropyelogram helps to delineate the ureter and pelvicalyceal system. Especially at the point of impaction of ureteric calculi, care must be taken to ensure the guidewire does not undermine the urothelium and perforate the ureter. The guidewire may need to be advanced past an impacted stone under direct vision by ureteroscopy.

For URS, the ureteric orifice is intubated. Rotating the ureteroscope under the guidewire allows the oval-tipped telescope to pass along the floor of the ureteric orifice with its major axis horizontal (**Figure 72.10**). For a difficult ureteric orifice, the hydrostatic pressure may be temporarily increased (after emptying the bladder), the ureteroscope can be passed over a second guidewire (**Figure 72.11**) or the ureteric orifice may be predilated by placement of a JJ stent for 10–14 days. For repetitive access to the proximal ureter or renal pelvis, deployment of a ureteric access sheath (internal diameter at least 2 Fr larger than the ureteroscope) can be considered in older children, although ureteric ischemia due to excessive stretch remains a concern.

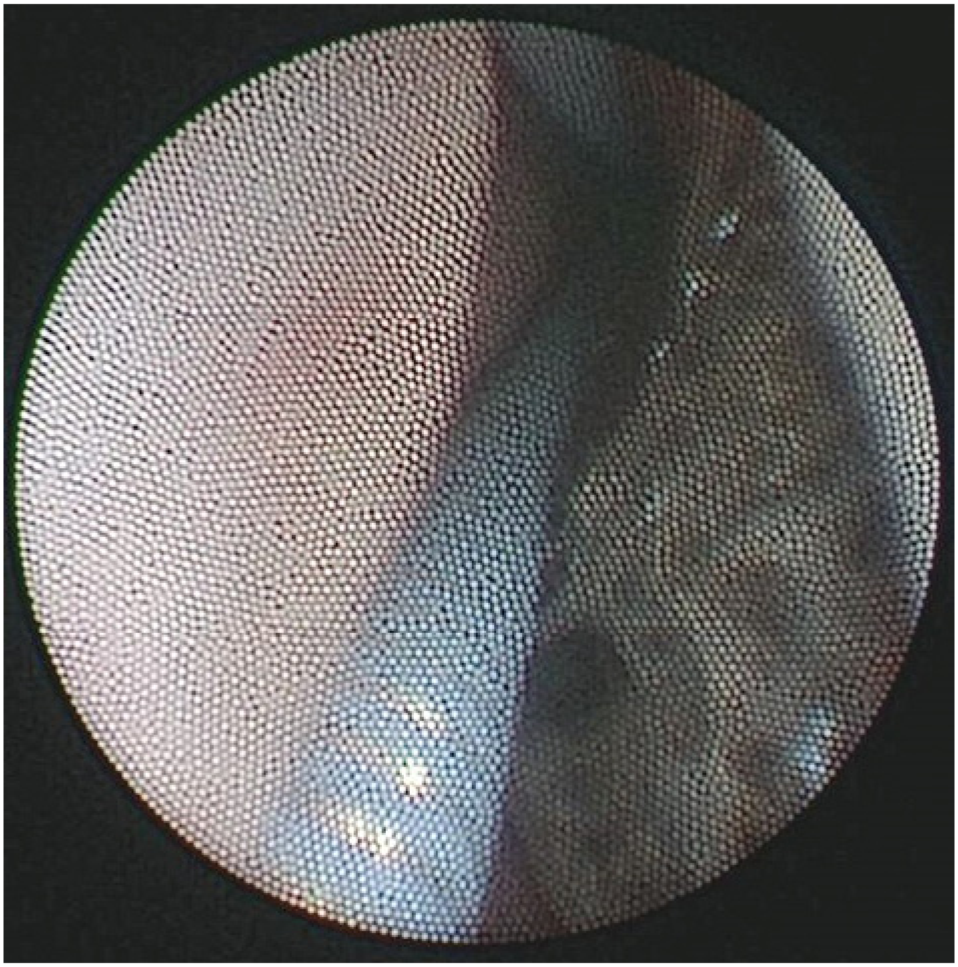


72.10



72.11 URS: With the patient in a lithotomy position, placement of a second guidewire helps to "open" the vesicoureteric junction/ureter to facilitate atraumatic passage of the ureteroscope.

The ureteroscope is carefully advanced while maintaining the ureteric lumen in the centre of the field of view and only in the absence of resistance (**Figure 72.12**). Holmium laser stone fragmentation within the pediatric ureter has a good safety profile and trauma to the urothelium can be minimized by placement of the laser fiber onto the stone surface. Slow irrigation or placement of a stone cone above the calculus will prevent retropulsion. Gentle passage of the scope above the stone for "filling" of the upper ureter maintains visibility at the stone with minimal irrigation. In addition, newer laser machines can generate a longer waveform, which permits stone dusting and again averts retropulsion.



72.12 URS: The image quality of the ureteric stone and two guidewires is compromised as a result of the very small caliber of the ureterosocpe and its optics.

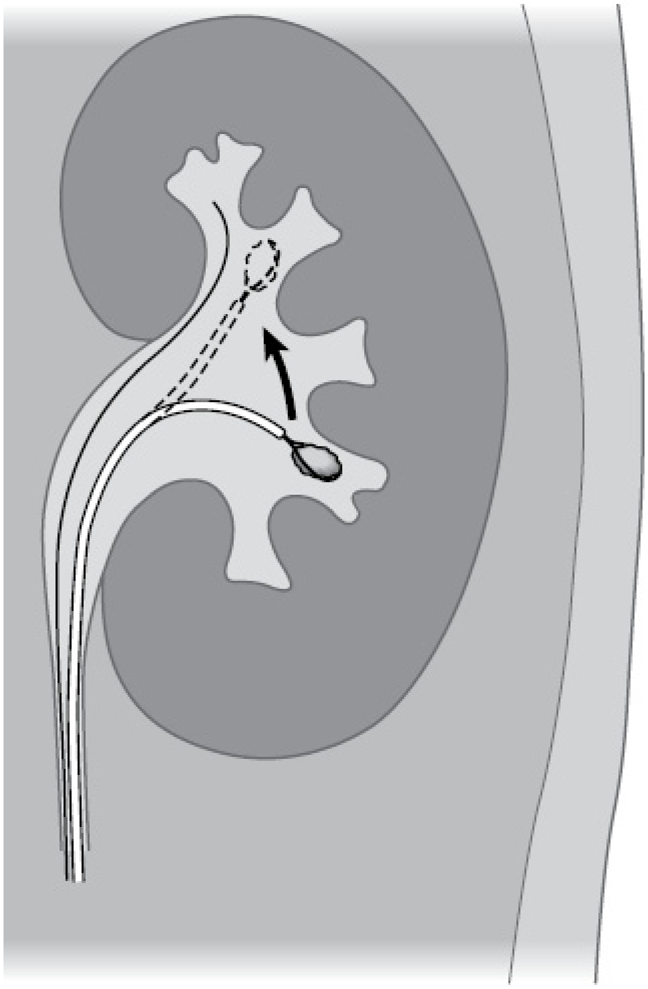
In the absence of ureteric or vesicoureteric junction (VUJ) oedema, stone fragments can be cleared under direct vision with a basket and sent for stone analysis. Very small stone fragments or stone dust may be left to pass spontaneously. The stone fragment for analysis must be sufficiently small to pass easily through the VUJ. A large fragment risks ureteric avulsion. By holding the stone close to the scope, and tilting its tip slightly upward through the VUJ, the passage of the basket and fragment are friction-free.

Placement of a JJ stent at the end of the procedure is advised in the presence of ureteric oedema (e.g. at the site of stone impaction), injury, or dilatation (e.g. after use of an access sheath), residual fragments, after recent infection, and in those with impaired renal function or a solitary or transplant kidney.

Retrograde intrarenal surgery

With continuing advances in flexible telescopes, renal stones are now accessible endoscopically in children. After initial inspection of the ureter by semi-rigid ureteroscopy, a ureteric access sheath may be placed over one of the two wires, to avoid trauma to the urethra and VUJ from repeated passages of scopes; alternatively, the flexible scope can be advanced over one of the soft-tipped wires under fluoroscopy into the kidney.

For RIRS, laser fibers and baskets must be inserted in the undeflected state to prevent damage to these expensive scopes. Lower-pole stones are best repositioned to an upper or interpolar calyx rather than lasered *in situ* (**Figure 72.13**). At maximal deflection, stone baskets and laser fibers cannot always be deployed, so stones in the lowermost calyces may remain inaccessible for retrograde endoscopic interventions. Retrograde access can also be used to laser incise strictures or open the neck to a calyceal diverticulum.



72.13

Minimally invasive cystolithotomy

Bladder stones can be dealt with in a variety of ways, including open cystolithotomy or by minimally invasive techniques either per urethra/per Mitrofanoff or percutaneously by PCCL. Endourological procedures afford a reduced length of stay and similar procedural time as open cystolithotomy with 90% stone clearance.

For PCCL, after suprapubic needle puncture under US or cystoscopic guidance, the tract is dilated over a guidewire, using first a disposable 9 Fr dilator followed by a NephroMax□ balloon (Boston Scientific, Marlborough, MA, USA) with an Amplatz sheath loaded. The balloon is partly deflated and the Amplatz advanced into the bladder, allowing telescopic visualization, fragmentation, and removal of the stone(s). PCCL avoids prolonged instrumentation of the urethra and decreases the risk of iatrogenic urethral stricture. The large working sheath facilitates the extraction of larger stone fragments and thereby reduces procedure time. After PCCL, a suprapubic catheter is left *in situ*, as well as a urethral catheter to enable irrigation if required.

In the presence of a Mitrofanoff channel, this can be gently dilated to an 18 Fr Amplatz for access onto a bladder stone without injury to the continence mechanism of the channel.

Laparoscopic operations

Depending on surgeon experience, laparoscopic stone removal can be considered for stones, for instance, located in a hydronephrotic renal pelvis needing pyeloplasty or in an abnormally located kidney. Reported stone-free rates range from 70% to 100% for laparoscopic pyelolithotomy and approach 100% for laparoscopic ureterolithotomy. Operative time for laparoscopic pyelolithotomy is almost double that of PCNL in comparative studies.

Open cystolithotomy

Although it remains a tool for large bladder stones, for ureteric and renal stones open surgery is now rarely indicated.

An incision is made in the lower abdomen to access the bladder. The bladder is opened and the stone removed. The bladder is closed with absorbable sutures, and a catheter is inserted into the bladder via the urethra.

POSTOPERATIVE CARE

In general, children are maintained on low-dose antibiotics until investigations are complete. Follow-up investigations include US to confirm complete absence of calculi, and MAG3 or DMSA. Surgery may occasionally be required to correct vesicoureteric reflux or an obstructive etiology. The risk of recurrent stones in children is as high as 50%. For children with no metabolic abnormality, who remain stone-free and have sterile urine for 2 years, recurrences are rare.

VIDEO

OPS_8e_video_72.1 Surgery for renal calculi

FURTHER READING

American Urological Association. *Surgical Management of Stones: AUA/Endourology Society Guideline (2016)*. Available at: https://www.auanet.org/guidelines/surgical-management-of-stones-(aua/endourological-society-guideline-2016)#x3164 [Accessed 15 June 2020.]

European Association of Urology. *Paediatric Urology*. Available at: https://uroweb.org/guideline/paediatric-urology/#3_14 [Accessed 15 June 2020.]

Guven S, Frattini A, Onal B, et al. Percutaneous nephrolithotomy in children in different age groups: Data from the Clinical Research Office of the Endourological Society (CROES) Percutaneous Nephrolithotomy Global Study. *Br J Urol Int* 2012; 111: 148–56.

Issler N, Dufek S, Kleta R, et al. Epidemiology of paediatric renal stone disease: A 22-year single centre experience in the UK. *BMC Nephrol* 2017; 18: 136.

Silay MS, Ellison JS, Tailly T, Caione P. Update on urinary stones in children: Current and future concepts in surgical treatment and shockwave lithotripsy. *Eur Urol Focus* 2017; 3(2–3): 164–71.

Smeulders N, Undre S. Minimally invasive interventions for stone disease. In: Godbole PP, Koyle MA, Wilcox DT (eds). *Pediatric Urology: Surgical Complications and Management*. Hoboken, NJ: John Wiley & Sons; 2015: pp. 125–31.

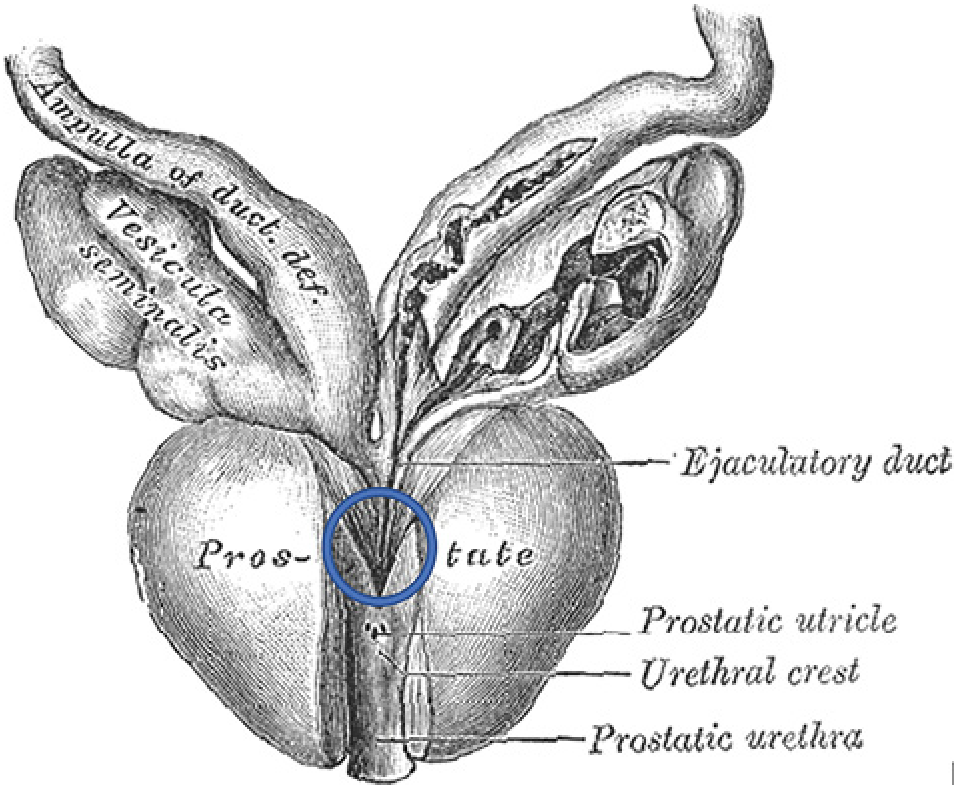
73

Posterior urethral valves

FARDOD O'KELLY AND MARTIN A. KOYLE

PRINCIPLES AND JUSTIFICATION

A posterior urethral valve (PUV), or more recently described as a congenital obstructive posterior urethral membrane (COPUM), is a structure that originates from the inferior margin of the verumontanum exclusively in males. Although its embryology remains uncertain (abnormal anterior fusing of plicae colliculi in posterior urethra; non-canalization of congenital urogenital remnants), it lies in the position of the infracollicular folds, which can often be discerned in the normal posterior urethra running downward from the verumontanum toward the bulb on either side of the midline (**Figure 73.1**).


Ampulla of duct. def.
Vesicula seminalis
Ejaculatory duct
Pros-
tate
Prostatic utricle
Urethral crest
Prostatic urethra

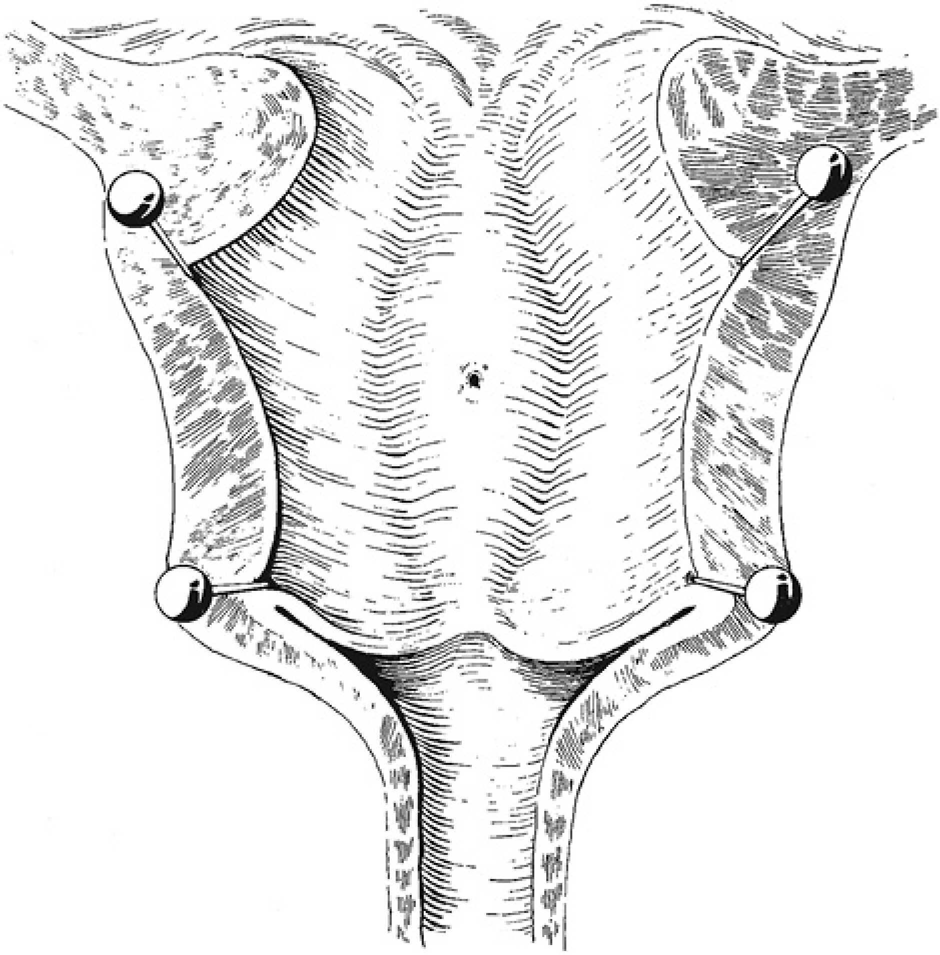
73.1 Coronal view of the posterior urethral with ejaculatory duct openings on either side of the prostatic utricle. The circle represents the site of origin of the PUVs.

The initial classification, described by Young in 1919, divided this condition into three entities:

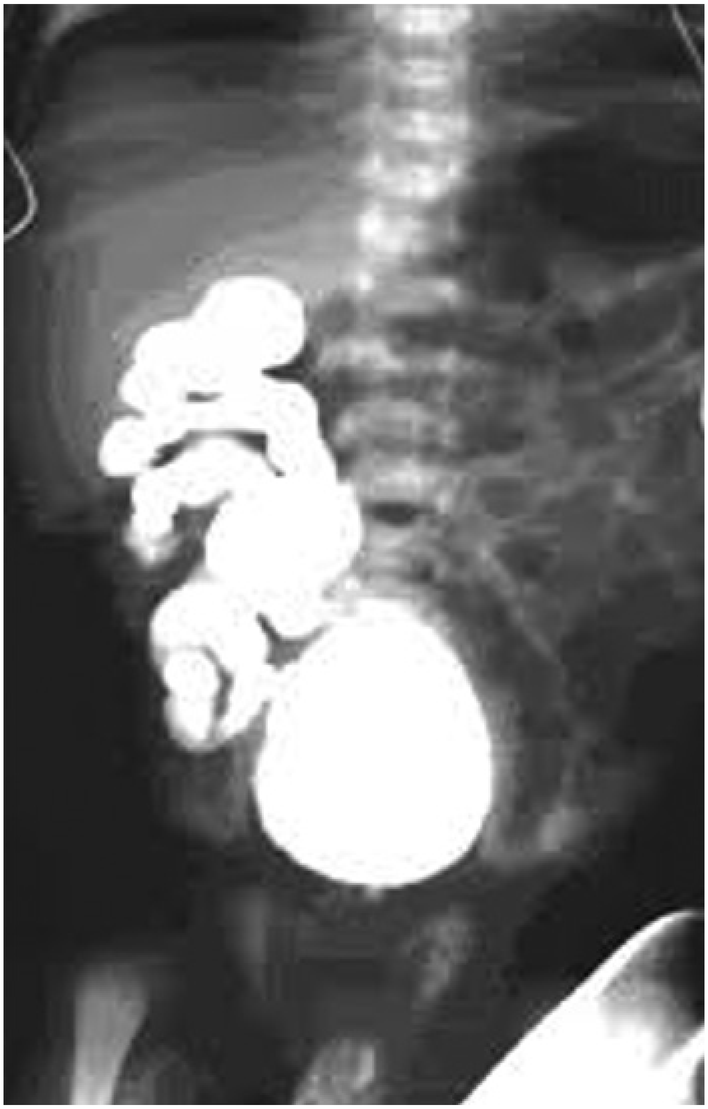
- *Type I:* Most common; extending from the bottom of the verumontanum distally along the prostatic and membranous urethra (95%)
- *Type II:* Longitudinal folds between the verumontanum and proximal prostatic urethra and bladder neck; no longer thought to be a distinct entity
- *Type III:* Disk of tissue distal to verumontanum (5%)

The incidence is approximately 1 : 5000 births, and, despite the advances in initial survival rates with early diagnosis and intervention, progression to end-stage renal disease still occurs in up to 50% of children by the age of puberty.

When exposed at autopsy through the anterior urethral wall, a PUV appears as two separate leaflets (**Figure 73.2**). It is unclear whether this represents either division of the membrane during postmortem dissection or *in vivo* membrane disruption during initial catheterization of the neonate, or whether it physiologically develops in this manner. Proximal to the valve "leaflets," there is barometric evidence of a dilated posterior urethra (crucial during radiological diagnosis), often a high and hypertrophic bladder neck, and bladder wall trabeculation with or without diverticulae and/or discrete areas of contained mucosal hemorrhage. The ureters may appear dilated and tortuous, and there may be hydronephrosis. This is usually bilateral, but may be asymmetric in the presence of vesicoureteric reflux with/without unilateral renal dysplasia (VURD) (**Figure 73.3**). This pop-off mechanism may also be demonstrated in the presence of calyceal rupture with urinary ascites.

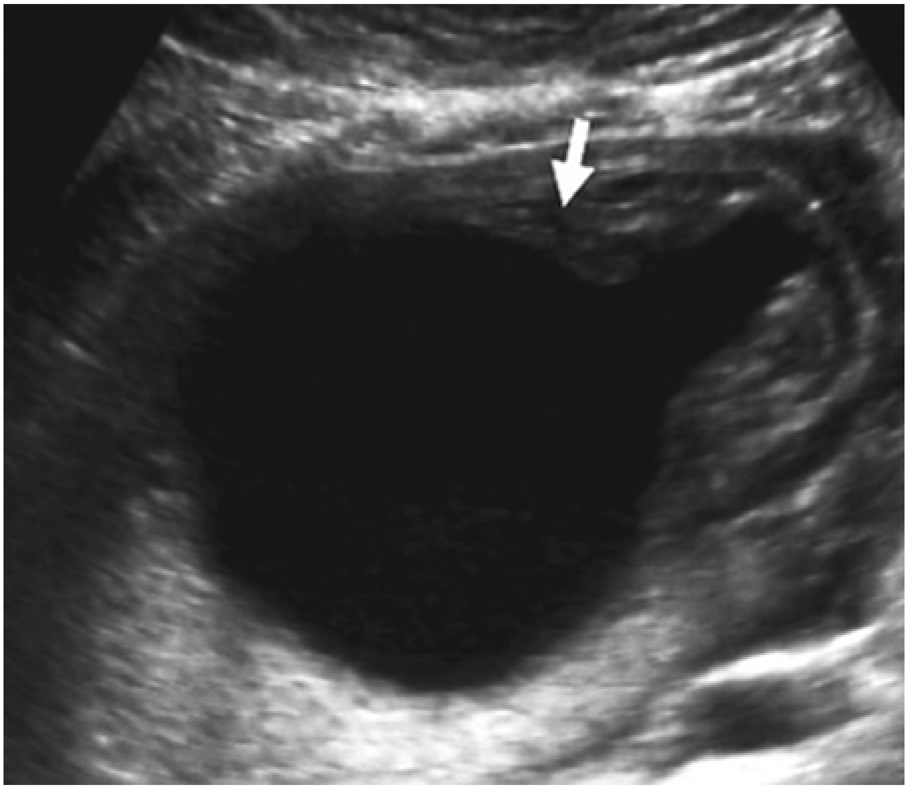


73.2



73.3

Diagnosis of PUV is often made prenatally in the contemporary setting. Prenatal diagnosis can be challenging with sonographic findings of increased bladder wall thickness and bladder dilatation felt to be the most accurate for correlation. The presence of a keyhole sign, while pathognomic, is not a reliable finding during sonography (**Figure 73.4**). Sonography is also useful for diagnosing dilated upper urinary tracts (hydroureteronephrosis).



73.4

In the postnatal period, the diagnosis of PUV may be made following clinical presentation with a palpable urinary, or an inability to void. In those children in whom a diagnosis is initially not made or missed, presentation may occur with respiratory difficulties (pulmonary hypoplasia), febrile urinary tract infection (UTI), unexplained fever with sonographic evidence of a congenital anomaly of the kidney and urinary tract, or with renal failure (acute/chronic). Children may demonstrate hyperkalemia, a raised nadir creatinine (>48 hours after birth), abnormal plasma osmolality, coagulopathy, and/or a severe metabolic acidosis. Boys presenting with PUVs after infancy may present with UTIs, poor urinary stream, voiding dysfunction, or daytime/night-time wetting. Creatinine appears to be the only independent risk factor for poor renal outcomes in PUV patients consistent with the literature. A finding of vesicoureteric reflux (VUR) is associated with the need for multiple surgeries but not with chronic kidney disease or end-stage renal disease.

PREOPERATIVE ASSESSMENT

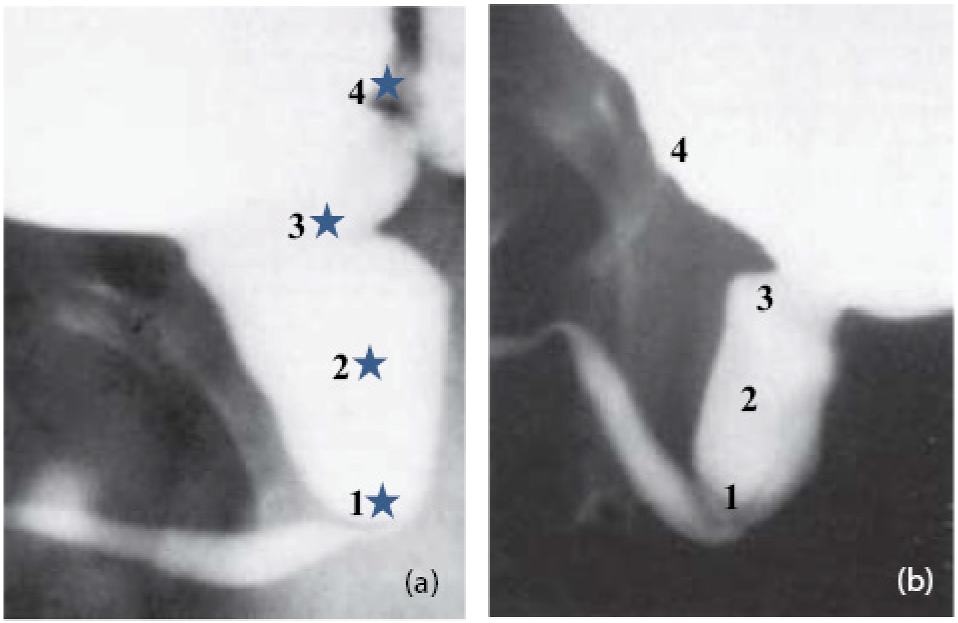
If a diagnosis of PUV is suspected prenatally, parents should be counseled carefully regarding the possibility of poor renal or respiratory function in the immediate postnatal period, the potential need for antibiotic prophylaxis, and the need for an early voiding cystourethrogram (VCUG), which may mandate emergent valve resection in the newborn period. Mothers should aim to deliver in a center with good neonatal capabilities with urological specialists on site or within easy transfer. On suspicion of the diagnosis, a 6–8 Fr plastic infant feeding tube should be passed transurethrally and secured for continuous bladder drainage. Self-retaining catheters should be avoided, as the hypertrophied bladder tends to clamp down around the balloon and obstruct the ureters. It may be necessary to use a curved (Coude-tipped) catheter in those with a difficult catheterization, as there may be coexistent bladder neck hypertrophy. It should be emphasized that iatrogenic urethral trauma is to be avoided. It is essential that the bladder drains well, and failure to do so is usually because the catheter has curled up in the dilated posterior urethra. Persistent difficulty can usually be resolved by injecting a few milliliters of contrast medium through the catheter and manipulating it under fluoroscopic control.

A full blood count including platelets, plasma electrolytes, creatinine, and acid–base status should be determined, and severe derangements, particularly hyperkalemia or a severe metabolic acidosis, should be corrected as a matter of urgency. An assessment should also be made of the infant's state of hydration. Pediatric nephrology advice should also be sought, not only in cases of renal impairment, but also as an introduction to the parents, as nephrology will continue to follow this child in the longer term.

When the urine appears to be infected (urinalysis) following a catheter-specimen sample, both a blood sample and a urine sample should be sent for culture, following which ampicillin and an aminoglycoside or a third-generation cephalosporin should be started intravenously. When septicemia is suspected, blood coagulation studies should also be carried out.

All infants with any respiratory distress should undergo chest radiography to exclude a pneumothorax secondary to pulmonary hypoplasia. In the majority of cases, the above actions will result in a rapid improvement in the infant's metabolic state and general condition. Those infants who remain in a toxic state or whose plasma creatinine does not begin to fall within 24 hours, despite correction of other metabolic abnormalities, would historically have been considered for percutaneous drainage of both kidneys, however this is now exceedingly rare in a contemporary setting.

The presence of a PUV should be confirmed by VCUG, but this should be delayed until urinary infection has been brought completely under control and metabolic disturbances have been corrected. A voiding film taken in the steep oblique projection during full micturition is necessary to demonstrate the valve. Dilatation of the urethra proximal to the valve is essential to the diagnosis (**Figure 73.5a**). Signs of bladder neck hypertrophy (**Figure 73.5b**) and bladder wall irregularity are also usually present. A very lax valve may occasionally prolapse down as far as the bulbar urethra. Here, the posterior run-off may not be readily apparent, but the filling defect caused by the valve leaflets can usually be made out running down from the verumontanum. It is also important to document the presence and grade of VUR, as well as the estimated cystographic bladder capacity.



73.5a,b

Most valves are thin, filmy structures that balloon downward during voiding, but a few are thicker and more rigid, forming a transverse obstruction in the mid-posterior urethra. As all true valves originate from the inferior aspect of the verumontanum, lesser degrees of valve severity are sometimes encountered, in which the two leaflets blend with the lateral urethral wall. They are unlikely to cause issues in the newborn period, but the effects they might have on voiding patterns in later childhood are unknown.

A variety of other conditions may masquerade as a PUV on the cystogram, and failure to recognize them often leads to inappropriate treatment. Among these are prominent infracollicular folds, which can sometimes be made out on a good-quality study in normal children. In these cases, however, the voiding phase of the cystourethrogram should demonstrate good passage of contrast into the anterior urethra with no posterior urethral dilatation.

Hesitant voiding in a normal baby may also cause an abrupt change in caliber of the posterior urethra, while extrinsic compression by the pelvic floor may cause one or more concentric indentations in the urethral contour. Neither of these is associated with evidence of obstruction above the lesion (dilated posterior urethra/bladder neck hypertrophy), however, and both should be regarded as normal variants. These normal variants are most often encountered during studies following episodes of febrile UTIs, where the index of suspicion for PUVs is low.

A neuropathic bladder may closely simulate a PUV, but the thin stream below the obstruction will be seen emerging from the center of the external urethral sphincter rather than from the posterior margin, as seen with a valve. In such cases, the spine should be carefully examined and other evidence sought of a neurologic deficit in the perineum or lower limbs. A posterior urethral stricture may cause a similar appearance, but this will usually be associated with a history of urethral or pelvic trauma. The cystographic appearances of prune-belly syndrome may mimic those seen with PUVs, but the correct diagnosis should be suspected from the appearance of the bladder, which lies horizontally and is invariably smooth-walled, and the dog-leg configuration of the posterior urethra, which often bears a utriculus masculinus. The clinical examination of the abdomen with/without cryptorchidism also lends a clear diagnosis.

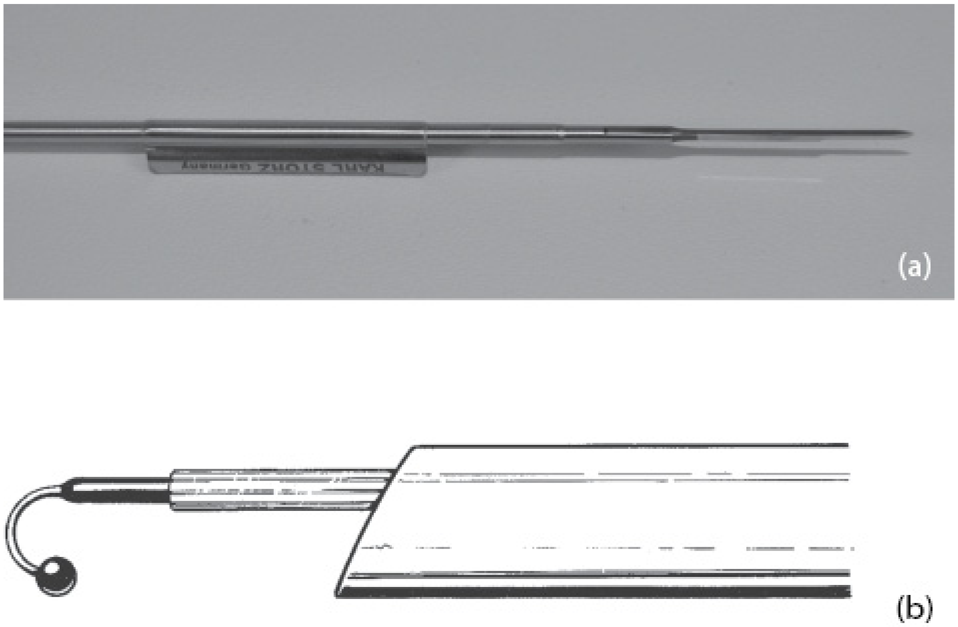
OPERATIONS

Resection in full-term infants and children

Under endotracheal anesthesia, the intubated infant is placed supine with the buttocks brought well down to the end of the operating table. The legs should be well protected with cotton wool and fixed with crepe bandages, usually in the frog-leg position (for neonates and infants), taking care to provide ample support to the thighs. Pediatric stirrups can be considered for larger children. In some cases the infant is very small, and bringing them to the end of the bed is not practical from an anesthetic perspective as they require access to the mouth and to IV lines, with consideration also given to tubing length. Therefore the infant can be placed perpendicular to the direction of the table, with the buttocks brought to the edge as described above. The skin is prepared with a topical povidone–iodine solution which is allowed to dry, and drapes are applied taking care to exclude the anus from the operative field. Fixing the posterior towel to the perineal skin with three 5/0 non-absorbable sutures (nylon/Prolene/silk) may be required to ensure that the anus does not become exposed during subsequent manipulations.

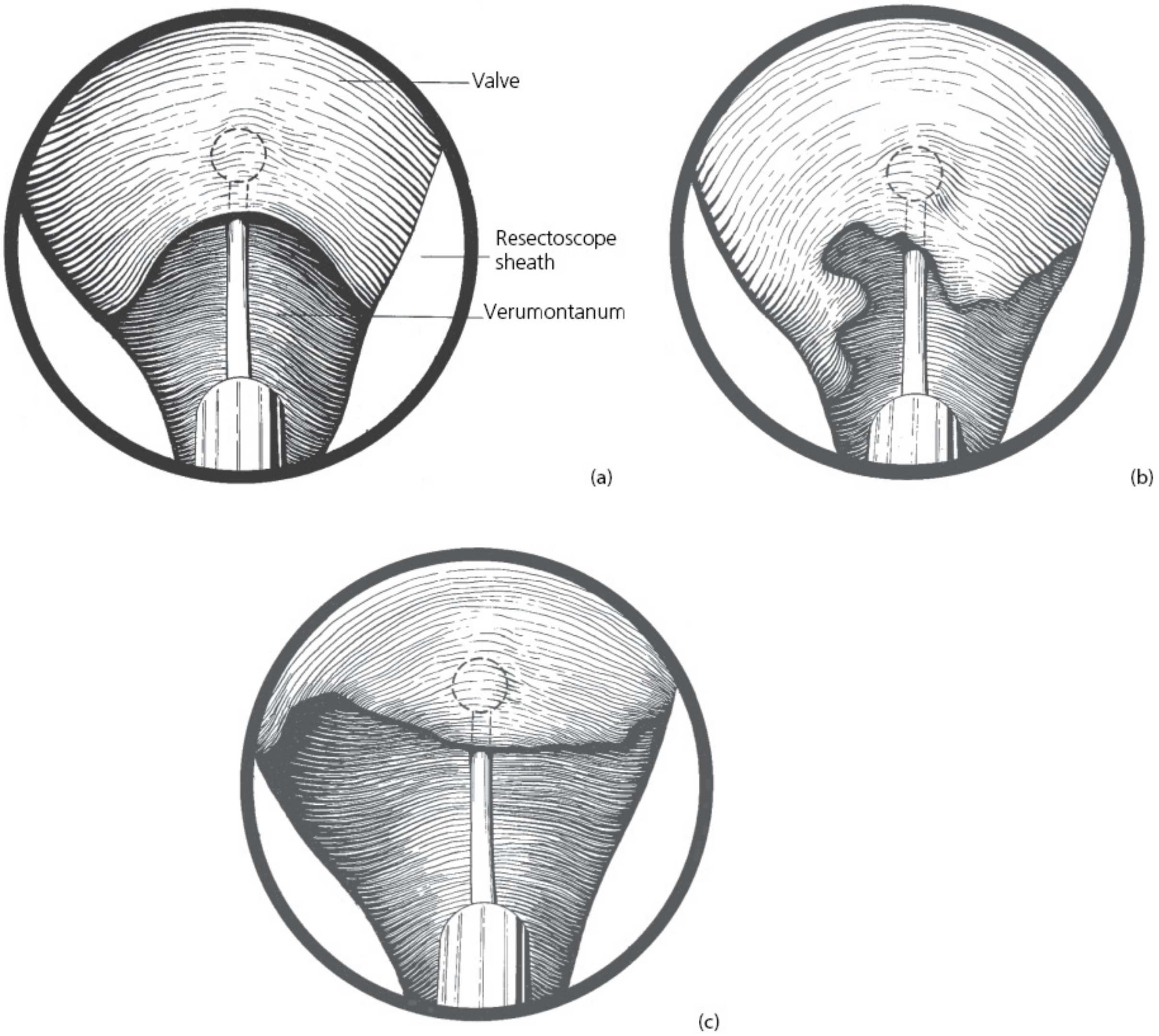
The caliber of the penile urethra should first be checked with a well-lubricated 8 Fr sound, which should be introduced only for 1–2 cm. If necessary, a meatotomy can be performed, but no attempt should be made to dilate the urethra. The diagnosis is then confirmed using a well-lubricated 6.5 mm or 9.5 Fr cystoscope introduced under vision. Retrograde passage of the cystoscope is carefully performed as the bladder neck may be hypertrophied and therefore quite high, requiring the surgeon to depress the scope to enter the bladder. Careful cystoscopic examination should be undertaken, noting the presence, degree, and location of trabeculation, diverticulae, and ureteric orifices. Passing the scope back in an antegrade direction will allow the surgeon to appreciate the verumontanum and the presence, quality, and thickness of valve leaflets in the 5 o'clock and 7 o'clock positions on the ventral surface of the urethra at the distal margin of the verumontanum, as the sharp outline of the inferolateral urethral walls are lost.

Resection of the valve can be undertaken using any number of attachments to the pediatric resectoscope and is dependent on experience, surgeon comfort, and local availability. Options include Sachse's cold knife urethrotomy (**Figure 73.6a**), hooked cold knife, hooked ball electrode (**Figure 73.6b**) (Karl Storz, Tuttlingen, Germany), and even holmium laser. The instrument is first assembled and the alignment of the working parts checked using the 0-degree telescope. The sheath is then dried and thoroughly coated with a water-soluble lubricant, and with its introducer in place is gently inserted through the meatus. The introducer is removed and the instrument reassembled and gently advanced under vision towards the bladder neck. It is usually necessary to angle the eyepiece end of the instrument downward to allow the beak to move anteriorly and pass through the bladder neck.



73.6a,b Resectoscope attachments used for the resection of PUVs. (a) Sachse's cold knife (Karl Storz); (b) hooked ball electrode.

Using a hooked ball electrode (see **Figure 73.6b**), the resectoscope once passed into the bladder is then rotated through 180 degrees (**Figure 73.7a**) as the resecting mechanism of most resectoscopes is situated in the superior midline. Irrigation fluid (generally water) should flow under low pressure, as the resectoscope is progressively withdrawn. Once through the bladder neck, the ball is run down along the anterior wall of the posterior urethra (**Figure 73.7b**) until just beyond the verumontanum where the valve suddenly snaps across the anterior portion of the field of view like a curtain. Further withdrawal of the instrument and manipulation of the trigger will cause the ball to engage the valve in the 12 o'clock position. A short burst of cutting current is then applied (**Figure 73.7c**).



73.7a–c

The procedure is very similar to that performed with a cold knife or hook. The resectoscope is returned to the normal position and advanced under vision back into the bladder. It is again rotated through 180 degrees and withdrawn into the posterior urethra to engage the now partially disrupted valve in the 12 o'clock position, where it is further disrupted. This maneuver should be repeated until it is certain that the anterior portion of the valve has been completely ablated. The instrument is again returned to the bladder and is rotated to engage residual valve tissue in the 5 o'clock and 7 o'clock positions. Any remaining freely floating tags do not require treatment. There is no objective evidence to suggest which directional combination of valve leaflet resections is the most effective.

The resectoscope is removed and the presence of an unobstructed urethra confirmed by manual expression of the bladder (Credé maneuver). Finally, an 8 Fr feeding tube is carefully passed, into the bladder. If resistance is encountered, a 0.9 mm (0.035 inch) polytetrafluoroethylene (PTFE) guidewire can be cystoscopically placed in the bladder and the catheter place over this following careful extraction of the cystoscope. This is to ensure that the catheter is not left curled up in the dilated posterior urethra. This catheter is then retained in place using a paper stitch or adhesive dressing (Tegaderm). There is no need to place a nylon suture through the prepuce or distal shaft skin. The catheter is then connected to a sealed drainage bag, and removed after 48 hours if the creatinine has reached a nadir, and there is no significant hematuria. If any significant bleeding occurs, attempts at valve ablation should be immediately discontinued and the situation reassessed after 2–3 days of catheter drainage.

There have been some studies exploring the role of bladder neck incision at the same time as PUV incision, but this is not yet widely performed, or endorsed by affiliated societies. Some data suggest that it is not associated with an additional risk of incontinence and dry ejaculation in early adulthood, and preserves antegrade ejaculation. At the current time, it is the authors' opinion that these data should be provided as an option but stress that they should still be considered experimental.

If there is evidence of dilating (high-grade) VUR on cystourethrography, then consideration should also be given to performing a circumcision at the same time as valve resection (method depends on surgical preference). This clearly needs to be discussed both with the parents, and with the anesthetic staff as it will likely mandate a longer anesthetic time.

Resection in preterm infants

It is inadvisable to attempt to pass a 9.5 Fr resectoscope in infants weighing less than 2.5 kg. In such cases, a few days of bladder drainage, using initially a 6 Fr urethral feeding tube, will have the effect of gently dilating the urethra so that a 6.5 Fr or even a 9.5 Fr resectoscope can be safely used. Alternatively, a very small cystoscope may be employed, introducing through the working channel a 3 Fr Bugbee electrode to coagulate the valve in a circumferential fashion.

Other techniques are also available for resecting the valve in a very small infant, but all have some disadvantages. The Whitaker hook electrode is a slender, insulated metal instrument that can be introduced through the urethra under fluoroscopic control and withdrawn to engage the valve. Short bursts of cutting current may relieve the obstruction, but the procedure is essentially blind and carries the risk of urethral trauma.

A Fogarty catheter with the balloon inflated with 0.1–0.3 mL of water has also been used to disrupt the valve, but this carries the risk of avulsion of the urethra and is now more of historical interest.

An alternative approach is to create a suprapubic cystotomy through which the valve can be resected in an antegrade fashion. Using a 10 Fr sheath and the hooked ball electrode, the valve is first engaged in the 12 o'clock position and coagulated. However, the hypertrophied bladder neck sometimes closes across the telescope lens so that the procedure may have to be carried out blindly. This is a much less common approach than using retrograde cystoscopic access.

Vesicostomy

In very small/preterm infants, another practical approach is to carry out a vesicostomy rather than attempt to disrupt the valve by the above methods. This is usually due to inadequate urethral diameter. A few months later, when the infant has reached an adequate size, the valve is coagulated through the urethra in the standard fashion. The vesicostomy is closed at the same time and the urethral catheter removed 10 days later. This allows the bladder to be kept decompressed and therefore acts as a protective mechanism for the upper tracts.

Upper tract drainage

Following relief of the urethral obstruction, correction of metabolic derangements, and eradication of infection, the plasma creatinine will in most cases rapidly fall to a nadir which is generally within the normal range for the patient's age. If it remains elevated, the possibility of obstruction of the dilated flaccid ureters as they pass through the hypertrophied bladder wall must be considered. In most cases, this phenomenon is transient and, provided that the infant remains well and the plasma creatinine is showing some improvement, an expectant policy may be adopted.

A persistently elevated or rising serum creatinine following successful valve ablation may be the result of infection, hyponatremia, or even antibiotic toxicity (aminoglycosides). However, when these causes have been excluded, percutaneous nephrostomies should be considered. A subsequent fall in the serum creatinine will confirm the diagnosis of obstruction at the level of the ureterovesical junction. In the case of percutaneous nephrostomy, after approximately 2 weeks, the tubes are clamped. If the serum creatinine remains low, they are removed. When this results in a rise of the serum creatinine, Sober Y cutaneous ureterostomies should be carried out to provide optimal tube-free upper tract drainage. The cutaneous limb can then be tied off when the infant is ready to come out of diapers. Alternatively, the ureters may be remodeled and reimplanted, but this operation is rendered difficult by young age (Paquin's law), the thickness of the bladder wall, and trabeculation, and in inexperienced hands complications are common. Failure of the serum creatinine to fall after placement of bilateral percutaneous nephrostomies is indicative of irreversible renal dysplasia and is a poor functional prognostic indicator.

POSTOPERATIVE CARE

Following removal of the urethral catheter, adequate emptying of the bladder should be confirmed clinically or by ultrasonography. Postoperative antibiotic prophylaxis is controversial in the absence of any further obstruction, and some units use cephalexein or trimethoprim–sulfamethoxozole (if over 3 months old), which is continued for 1 month. The presence of VUR should not necessarily alter postoperative management and depends on the findings. If a pop-off mechanism is encountered with a unilateral VURD, no prophylaxis is required. If the child is circumcised, or has non-dilating reflux (Grade 1–2), there is also no strong objective evidence for antibiotic prophylaxis.

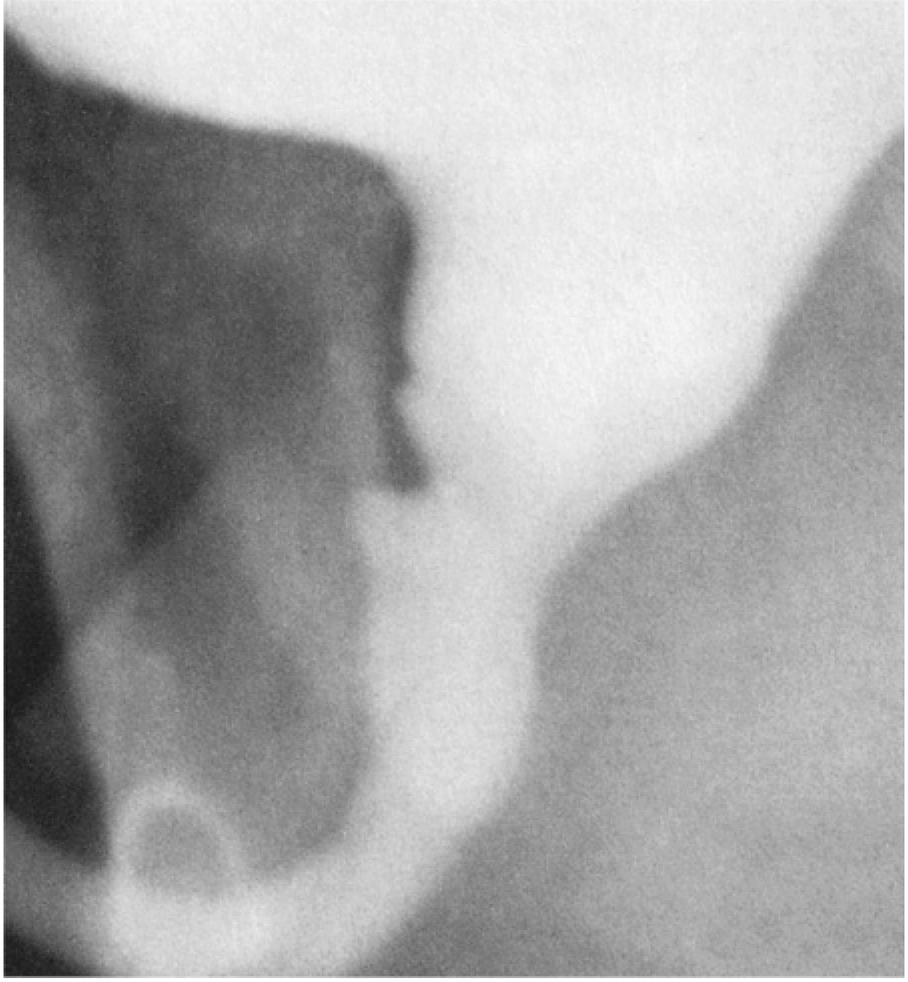
Sodium bicarbonate supplements may also often be necessary to correct a persistent metabolic acidosis, and these may need to be given for a year or more. Polyuria is also common, and the parents should be advised to give supplementary clear feeds early in the event of a diarrheal illness.

The role of clean intermittent catheterization in those boys who have undergone valve ablation remains controversial, but it should be considered in those who are unable to adequately empty their bladders autonomously. In those cases, overnight drainage should also be considered. There are data in the latter that overnight drainage reduces diuresis, reduces the frequency of UTIs, improves upper tract dilatation, and improves continence. Overnight drainage in conjunction with daytime clean intermittent catheterization may prove to be an alternative to bladder augmentation in select children with poorly compliant bladders and early stages of renal compromise. Other studies have suggested a role for the creation of a Mitrofanoff stoma, and that this can lead to significant improvements in hydronephrosis and urodynamic bladder dysfunction parameters. However, it does not prevent renal deterioration.

At 3 months, measurement of the split function and drainage of each kidney may be considered using a frusemide ^{99m}Tc-MAG3 (mercaptoacetyl triglycine) scan. This will act as a baseline for any future studies. A blood sample is also taken to check the plasma creatinine, electrolytes, and acid–base status.

At approximately 6–8 weeks, an interval VCUG is repeated for quality control to confirm adequate resection of the valve and the absence of any stricture of the urethra, and to determine whether any previously noted VUR is still present.

In about one-third of cases, it will be found to have disappeared (**Figure 73.8**). An interval ultrasound (US) is also performed at 3 months to assess the appearance of the upper urinary tracts. If there is a sonographic suggestion of renal scarring in the presence of persistent VUR, then a ^{99m}Tc-DMSA (dimercaptosuccinic acid) static renogram may be considered to confirm this and to determine the contribution of the kidney on the refluxing side to total renal function.



73.8

UTIs occurring during childhood after successful valve ablation are often due to incomplete bladder emptying. Double or triple micturition should be tried before instituting clean intermittent catheterizations.

All infants and those older children with impaired renal function at presentation will require close supervision until they reach adulthood. A progressive rise in plasma creatinine is often seen during childhood, and in the most severe cases, renal transplantation may be required before puberty. Persistent urinary incontinence is an indication for cystometrography (pressure-flow urodynamics). Bladders demonstrating very poor compliance may require augmentation, which should be carried out before transplantation. However, data have also demonstrated that augmentation cystoplasty can increases the risk of UTI after renal transplantation and that pre-emptive augmentation cystosplasty should be constructed only if the risks associated with increased bladder pressures exceed those following augmentation.

Since a considerable number of PUV patients with high-pressure bladders eventually develop myogenic failure, some units advocate postponing augmentation in this population, as long as they are under close surveillance. Other data would suggest that the outcomes of renal transplantation in PUV patients are similar to those undergoing a renal transplant without an obstructive uropathy, and therefore bladder dysfunction may not have a major role in affecting transplant graft survival.

FURTHER READING

Bernardes LS, Aksnes G, Saada J, et al. Keyhole sign: How specific is it for the diagnosis of posterior urethral valves? *Ultrasound Obstet Gynecol* 2009; 34(4): 419–23.

Bilgutay AN, Roth DR, Gonzales ET Jr, et al. Posterior urethral valves: Risk factors for progression to renal failure. *J Pediatr Urol* 2016; 12(3): 179.e1–7.

Casella DP, Tomaszewski JJ, Ost MC. Posterior urethral valves: Renal failure and prenatal treatment. *Int J Nephrol* 2012; 2012: 351067.

Chertin B, Cozzi D, Puri P. Long-term results of primary avulsion of posterior urethral valves using a Fogarty balloon catheter. *J Urol* 2002; 168: 1841–3.

Clifton MS, Harrison MR, Ball R, Lee H. Fetoscopic transuterine release of posterior urethral valves: A new technique. *Fetal Diagn Ther* 2008; 23: 89–94.

Dewan PA, Keenan RJ, Morris LL, Le Quesne GW. Congenital urethral obstruction: Cobb's collar or prolapsed congenital obstructive posterior urethral membrane (COPUM). *Br J Urol* 1994; 73(1): 91–5.

Ghanem MA, Nijman RJ. Long-term follow up of bilateral high (Sober) urinary diversion in patients with posterior urethral valves and its effect on bladder function. *J Urol* 2005; 173: 1721–4.

Hebenstreit D, Csaicsich D, Hebenstreit K, et al. Long-term outcome of pediatric renal transplantation in boys with posterior urethral valves. *J Pediatr Surg* 2018; 53(11): 2256–60.

Heikkila J, Rintala R, Taskinen S. Vesicoureteral reflux in conjunction with posterior urethral valves. *J Urol* 2009; 184: 1555–60.

Jesus LE, Pippi Salle JL. Pre-transplant management of valve bladder: A critical literature review. *J Pediatr Urol* 2015; 11(1): 5–11.

Keihani S, Kajbafzadeh AM, Kameli SM, Abbasioun R. Long-term impacts of concurrent posterior urethral valve ablation and bladder neck incision on urinary continence and ejaculation. *Urology* 2017; 99: 278–80.

King T, Coleman R, Parashar K. Mitrofanoff for valve bladder syndrome: Effect on urinary tract and renal function. *J Urol* 2014; 191(5 Suppl): 1517–22.

Kousidis G, Thomas DF, Morgan H, et al. Long-term outcome of prenatally diagnosed posterior urethral valves: A 10- to 23-year follow-up study. *BJU Int* 2008; 102: 1020–4.

Mandal S, Goel A, Kumar M, et al. Use of holmium:YAG laser in posterior urethral valves: Another method of fulguration. *J Pediatr Urol* 2013; 9(6 Pt B): 1093–7.

Nguyen MT, Pavlock CL, Zderic SA, et al. Overnight catheter drainage in children with poorly compliant bladders improves post-obstructive diuresis and urinary incontinence. *J Urol* 2005; 174(4 Pt 2): 1633–6.

Stuhldreier G, Schweitzer P, Hacker HW, Barthlen W. Laser resection of posterior urethral valves. *Pediatr Surg Int* 2001; 17: 16–20.

74

Hypospadias

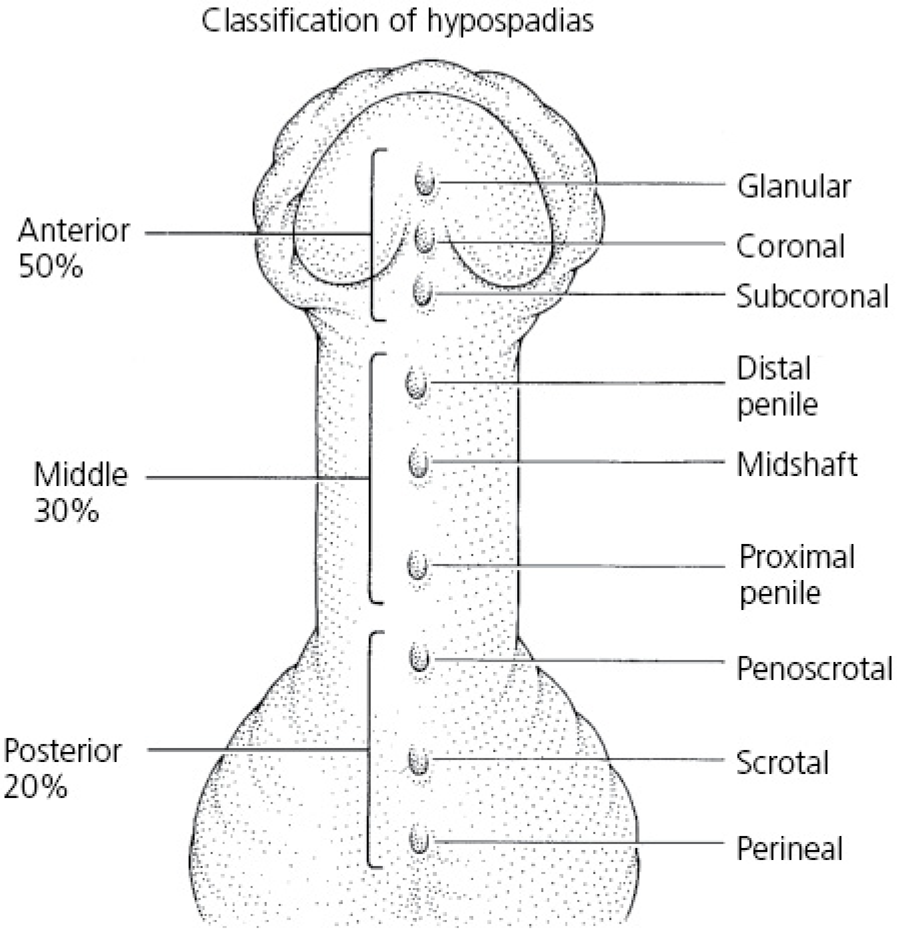
BRYAN S. SACK AND JOHN M. PARK

INTRODUCTION

Hypospadias is classically defined as an association of three congenital anatomic anomalies of the penis:

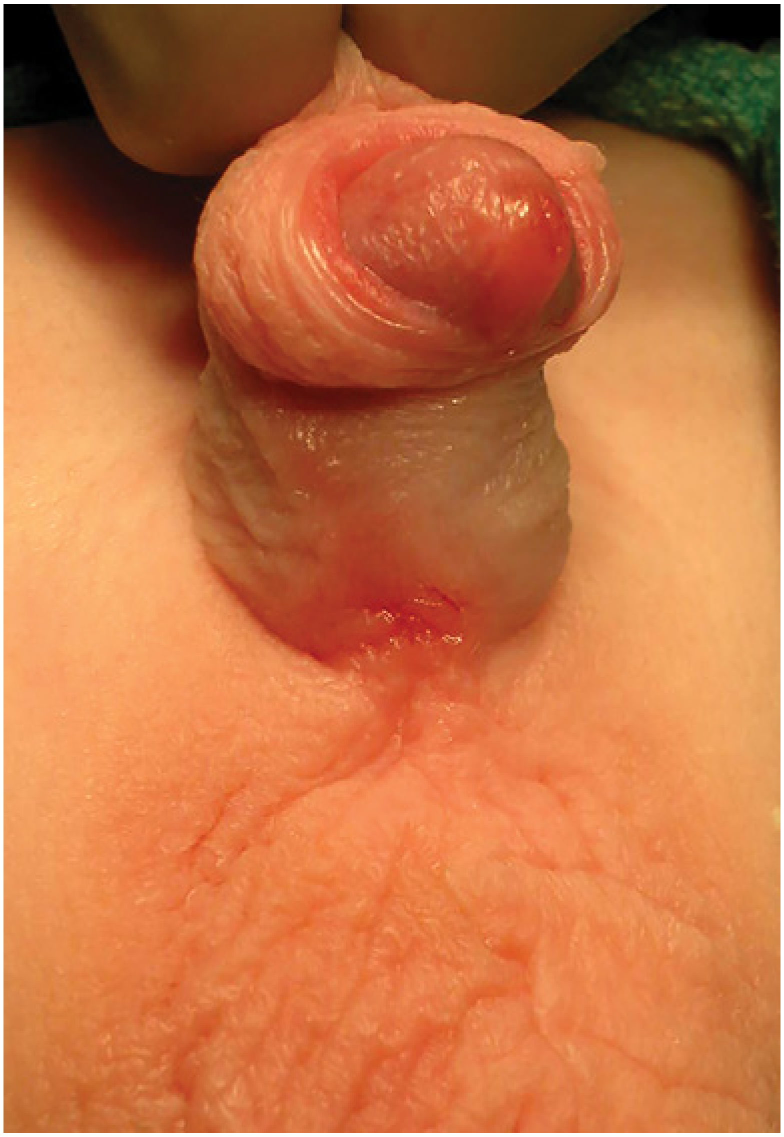
- An abnormal ventral opening of the urethral meatus
- An abnormal ventral curvature of the penis (called "chordee")
- An abnormal dorsal "hood" distribution of the foreskin with ventral deficiency

The diagnosis of hypospadias is evident on newborn examination, but in some instances of mild hypospadias, such as the megameatus-intact prepuce (MIP) variant, in which the foreskin is normal, detection of hypospadias might not occur until the foreskin is retracted. The most common classification system employs anatomic description of meatal position (**Figure 74.1**).

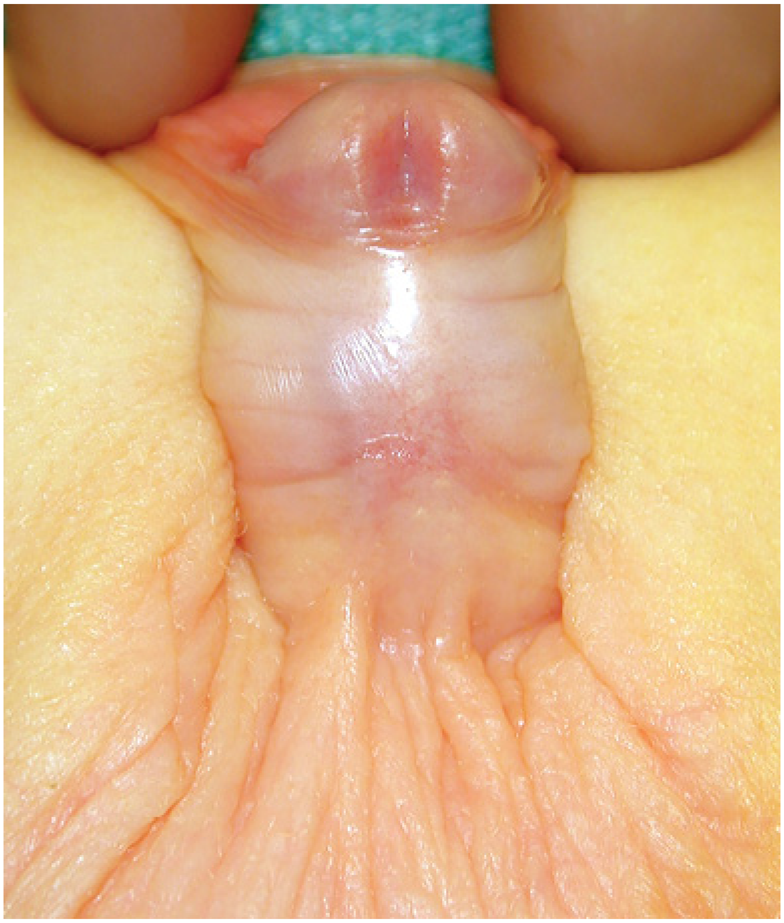

Classification of hypospadias
Anterior 50%
Middle 30%
Posterior 20%
Glanular
Coronal
Subcoronal
Distal penile
Midshaft
Proximal penile
Penoscrotal
Scrotal
Perineal

74.1

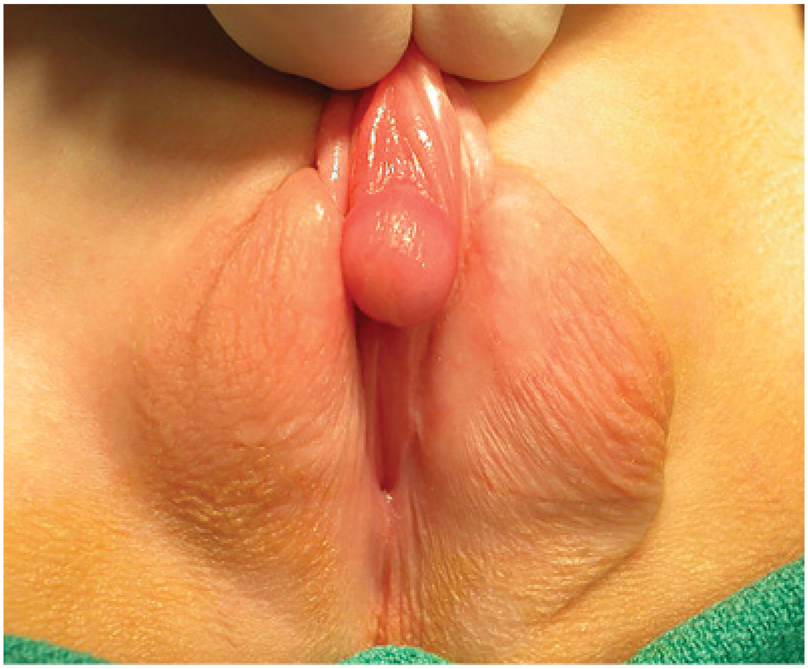
Glanular, coronal, subcoronal and distal penile positions constitute the majority of cases (approximately 70%), but hypospadias can occur in more severe forms with the urethral meatus opening near the scrotum and perineum (**Figures 74.2–74.4**).



74.2 Mild hypospadias in which the urethral meatus is distally located and the penile shaft is associated with mild chordee and ventrally deficient prepuce.



74.3 Moderate hypospadias in which the urethral meatus is subcoronal. There is a deep glanular groove distal to the meatus (called the urethral plate), and the prepuce is deficient ventrally.



74.4 Severe hypospadias with the urethral opening in the perineum.

INCIDENCE AND ASSOCIATED CONDITIONS

Hypospadias has been reported to occur approximately in one out of 100–300 live male births. In a case-controlled study by Sweet et al., hypospadias was present once in every 122 births. The great majority (87%) were mild—coronal or glanular. Interestingly, the rate of severe hypospadias seems to have increased three- to fivefold over the last few decades. This trend could reflect more frequent reporting and earlier diagnosis, or it may reflect other undefined biological factors, such as environmental endocrine disruptors during pregnancy. Hypospadias is most likely an inherited congenital defect, as it occurs in 6–8% of fathers of affected boys and 14% of male siblings.

Associated anomalies include cryptorchidism (8–9%) and inguinal hernias/hydrocele (9–16%). The rate of associated genital anomalies increases in patients with more severe proximal defects. A disorder of sex development should be considered in patients with concomitant hypospadias and cryptorchidism. This association is even stronger with bilateral cryptorchidism. Patients with severe hypospadias may also have a significantly enlarged utricle, which may serve as a nidus for urinary tract infections (UTIs), cause issues with catheterization at the time of reconstruction, and cause future voiding difficulties.

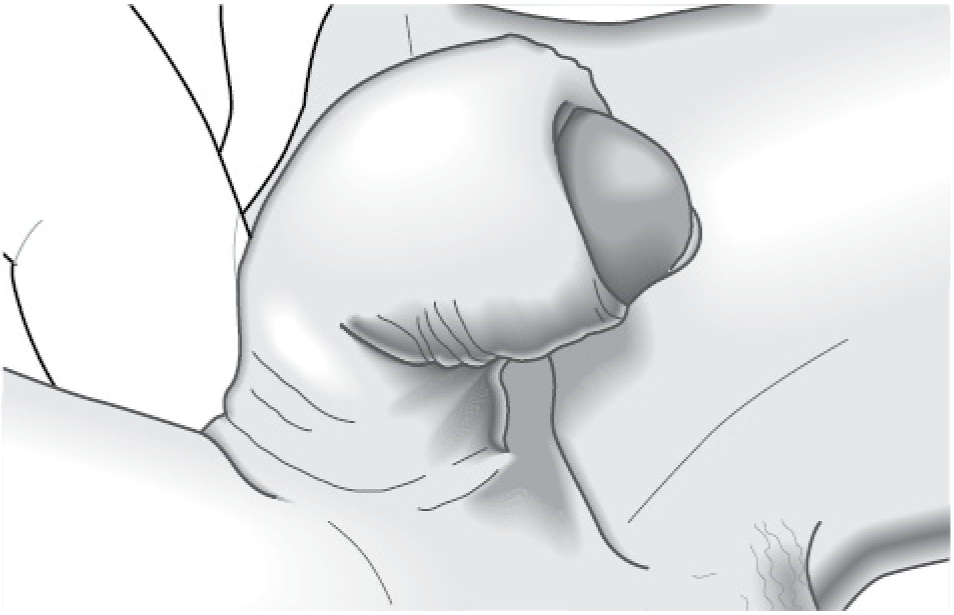
Chordee (congenital penile curvature)

Early on, the ventral curvature was thought to be caused primarily by fibrous tissues encasing the hypospadiac urethra, thereby tethering and bowing the penis, and this led to the proposal that the most important part of the chordee correction was aggressive excision of so-called "chordee tissues." Penile embryology and anatomic studies have shown that the etiology of chordee is much more complex, and it includes:

- Abnormal development of the urethral plate (the mucosa-lined longitudinal groove between urethral meatus and glans)
- Abnormal, fibrous tissue at the urethral meatus
- Corporal disproportion between dorsal and ventral cavernosal tissues

The concept of corporal disproportion is particularly important in that a routine division and lifting of the urethral plate should no longer be considered as the principal maneuver in chordee correction. Indeed, the preservation of the urethral plate for neourethra reconstruction along with corporal plication for chordee correction constitutes a fundamental approach in modern hypospadias repair techniques.

Although chordee is commonly seen in association with hypospadias, it can occur in isolation with orthotopically located urethral meatus (Figure 74.5). In some cases, this is caused by a simple skin tethering, representing a simple surgical problem. In other instances the chordee is a "form frusta" of hypospadias in that more extensive anatomic abnormalities coexist such as thin, dysplastic urethra and disproportionate corpora, which may require a significant reconstruction.



74.5

Megameatus-intact prepuce

In this situation, the prepuce is circumferentially formed and the hypospadiac urethral meatus is not evident until the foreskin is fully retracted (i.e. at the time of circumcision). Typically, the meatus is at or just distal to the corona with a wide and deep urethral plate groove that can be tubularized into the neourethra. In most situations, MIP is not associated with significant chordee.

PREOPERATIVE ASSESSMENT AND PREPARATION

Preoperative evaluations

In general, the literature does not support routine evaluation of the urinary tract with either ultrasonography or other imaging modalities for hypospadias patients. In patients with a severe defect, such as perineal hypospadias, lower urinary tract evaluation with either endoscopy (at the time of repair) or voiding cystourethrography may define the utricle size and reveal concurrent anomalies that may aid in surgical planning and postoperative management. Routine endocrinologic and genetic evaluation of patients with isolated hypospadias is probably unnecessary, unless the defect is severe or associated with cryptorchidism. In instances of concomitant cryptorchidism, disorders of sexual development have been seen in up to 15% of unilateral and 30% of bilateral cases.

Hormonal manipulations

There is considerable disagreement regarding the use of hormonal stimulation prior to hypospadias repair for the purpose of penile enlargement. Suggestions were made that preoperative human chorionic gonadotropin treatment may decrease the severity of hypospadias and increase the vascularity and thickness of proximal corpus spongiosum, thereby allowing more simple repairs. Preoperative testosterone was shown to increase penile size, along with improved skin availability and local vascularity. Hormonal supplementation may be useful in children with microphallus and with previously failed repairs, in which local tissues are deficient. However, multiple retrospective reviews have failed to demonstrate a decrease in complications or appearance of the reconstructed phallus with administration of testosterone preoperatively.

Timing of surgery

A report by the American Academy of Pediatrics suggested that the best time for hypospadias surgery may be between 6 and 12 months of age. This recommendation was made based on a number of factors, including the psychological effects of the genital surgery in children, improved technical aspects of hypospadias surgery, and advances in pediatric anesthesia. The risk of neurotoxicity from general anesthesia in infants continues to be researched and the age recommendation of performing non-urgent procedures may change in the future. Postoperative management issues such as the care of neourethral catheter and wound dressings are also significantly easier and safer in infants.

Anesthesia

In young children, general anesthesia with endotracheal intubation is the most reliable approach. Adjunctive regional analgesia using long-acting injectable nerve-blocking agents, such as bupivacaine, delivered via either caudal route or penile block, is safe and efficacious. To obviate the risk of general anesthesia, spinal anesthesia alone for distal repairs is starting to be used in the infant cohort.

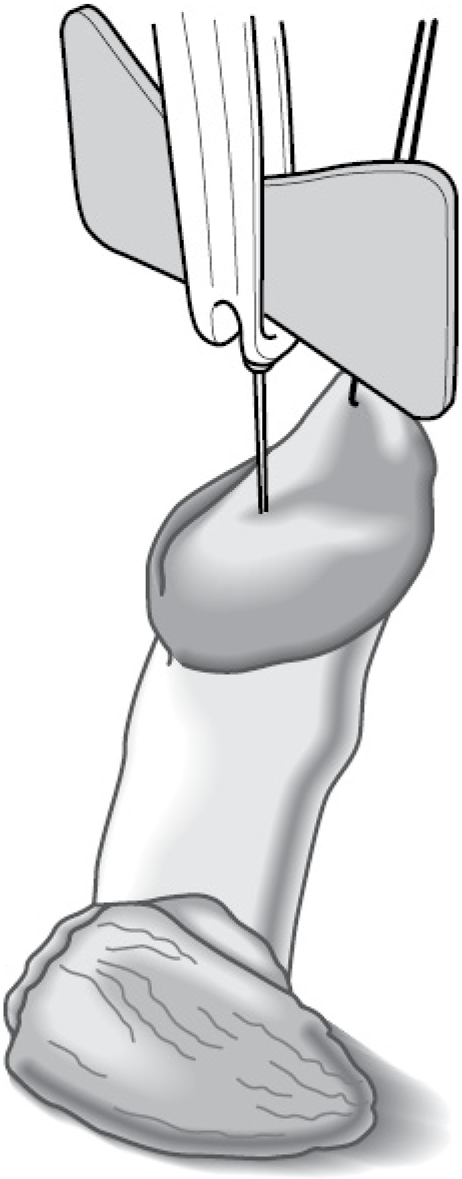
SURGICAL PRINCIPLES

Routine perioperative antibiotics are not necessary, unless the urethra and bladder are intubated postoperatively. Hemostasis must be accomplished with precision, applying judicious cauterization, since ischemic tissue necrosis will lead to the breakdown of repair, infection, and urethrocutaneous fistula. Careful use of vasoconstrictive agent (epinephrine diluted to 1 : 200 000) around the region of the corona and glans will also reduce the amount of bleeding intraoperatively. A tourniquet may be applied to the base of the penis during certain parts of the surgery, but it must not be used excessively for a prolonged time. Most surgeons rely on optical magnification to enhance surgical precision.

Orthoplasty (chordee correction)

The degree of ventral curvature influences the type of hypospadias repair. Even if the urethral opening is not severely ectopic, the penis may require an extensive reconstruction (even a staged one) in order to accomplish adequate straightening of the phallus. Preoperatively, gentle retraction of the penile base allows a reasonable assessment of the location and the severity of overall chordee.

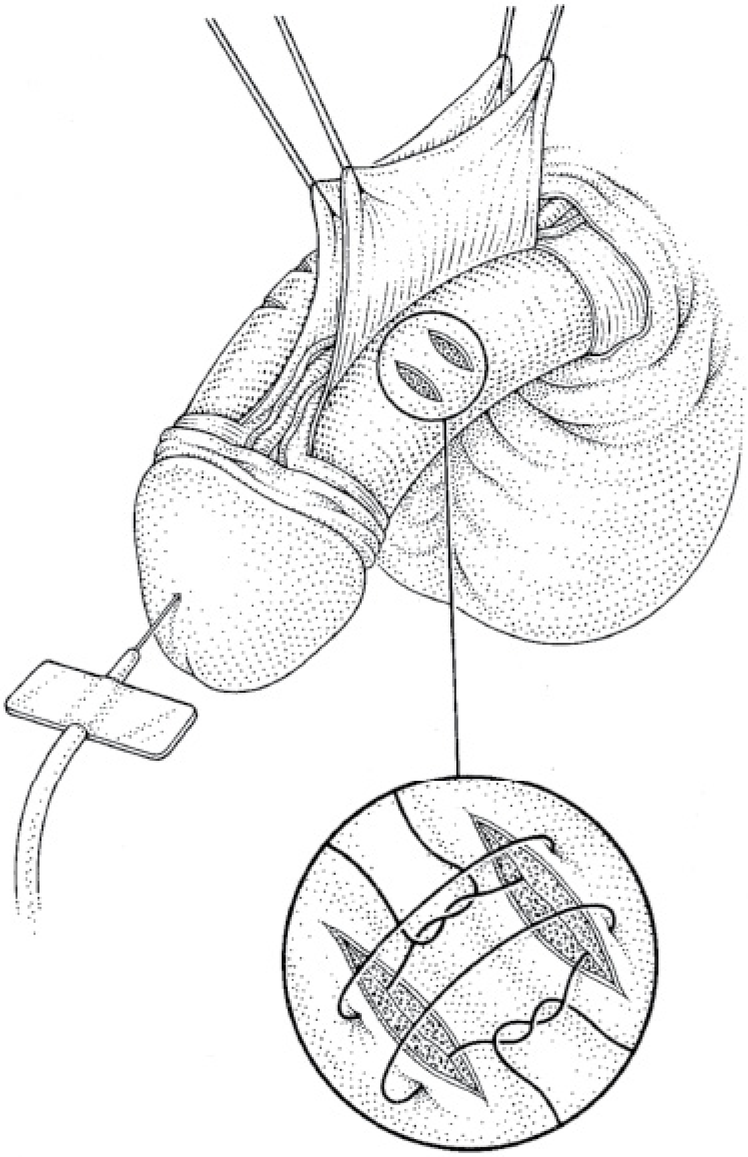
Intraoperative evaluation of chordee using an artificial erection test is critical (**Figure 74.6**). After the degloving of the penile shaft, injection of one of the corpus cavernosum is performed directly using 5–10 mL of saline solution with a small 23-gauge butterfly needle while a penile tourniquet is in place at the base of the penis.



74.6

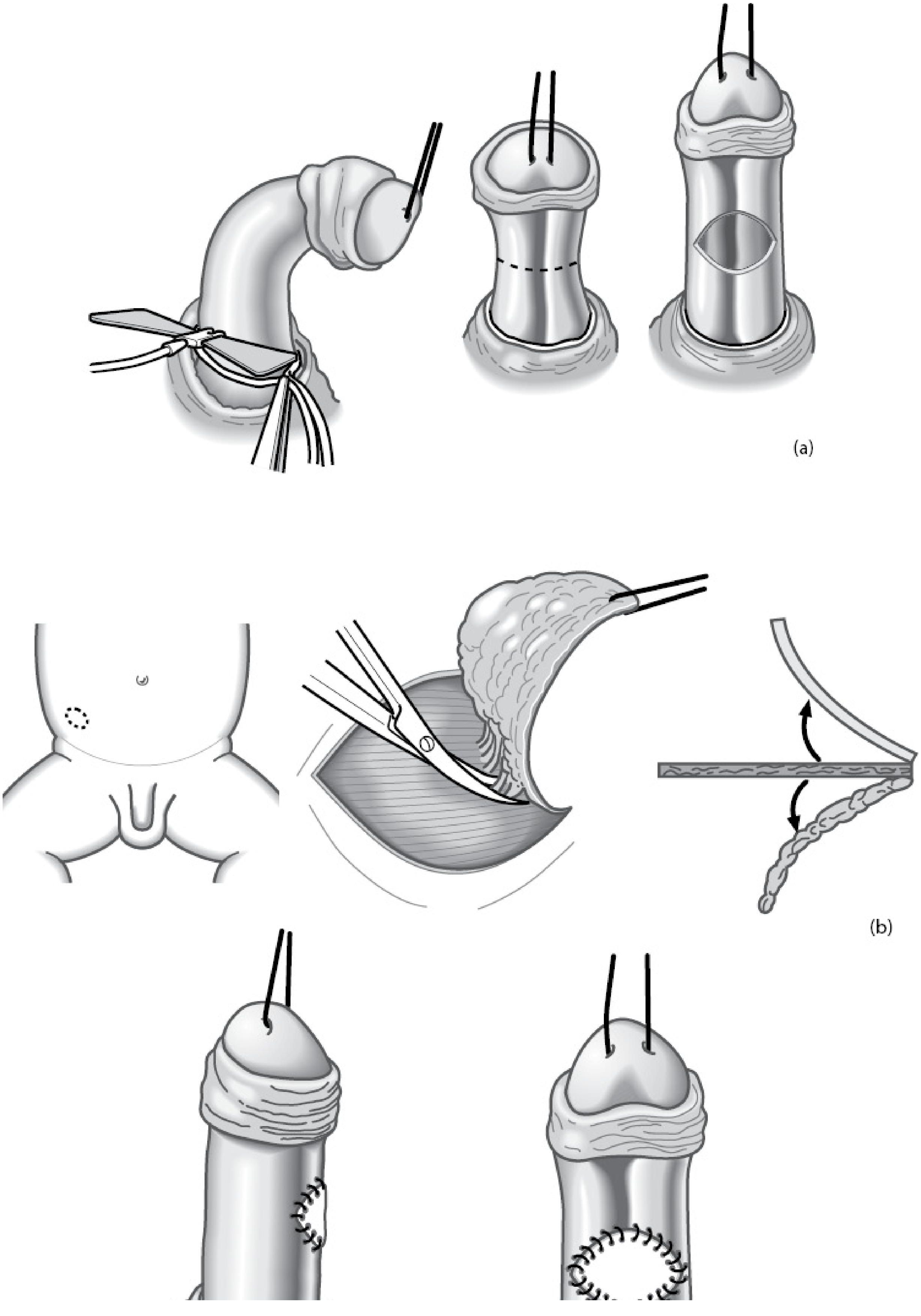
At times, release of tethering ventral skin may be all that is required for orthoplasty. In this situation, some of the dorsal prepuce and penile skin may need to be transposed ventrally to make up for the skin deficiency.

One of the most commonly used orthoplasty techniques is dorsal corporal plication, as described by Nesbit. His original description involved mobilization of the dorsal midline neurovascular bundle, followed by making an elliptical excision of the dorsal tunica albuginea vertically and approximating these defects horizontally (the Heineke–Mikulicz principle) (**Figure 74.7**). This concept was extended further by Baskin and Duckett, in which two parallel lines of incision are made in the tunica albuginea transversely on the dorsolateral aspects of each corpus cavernosum at the point of maximal curvature, and the outer edges of the incisions are approximated using buried sutures. This technique is reserved for mild degrees of chordee (<30 degrees) and when plication will not cause significant penile shortening.



74.7

In patients with severe curvature or with short phallic length, dorsal plication technique may not be optimal because of potential shortening, and ventral transverse incision of tunica albuginea and patch grafting may be more suitable (**Figure 74.8b**). The corporal defect created after the incision is then covered with de-epithelialized dermis harvested from the lower abdominal wall, tunica vaginalis, or a synthetic graft in older patients (**Figure 74.8b**). At the level of greatest curvature, a deep transverse incision is made ventrally, exposing the cavernosal tissue. This incision must extend from one lateral corner to the other to fully correct the curvature. Preferably, a de-epithelialized elliptical patch of dermis from the inguinal crease is sewn to the tunica albuginea edges using a running suture (**Figure 74.8c**). Obtaining a rigid artificial erection after ventral grafting to assess residual chordee is often difficult and emphasizes the need for aggressive corporal incision to ensure complete correction.


(a)
(b)

74.8a–c

Hypospadiac penis maybe associated with penile torsion, most often in the counterclockwise direction. In mild cases, this may be corrected simply by rotating the penile skin coverage, but in more severe cases, additional correction may be required, such as corporal body tacking and dorsal dartos flap rotation.

Urethroplasty

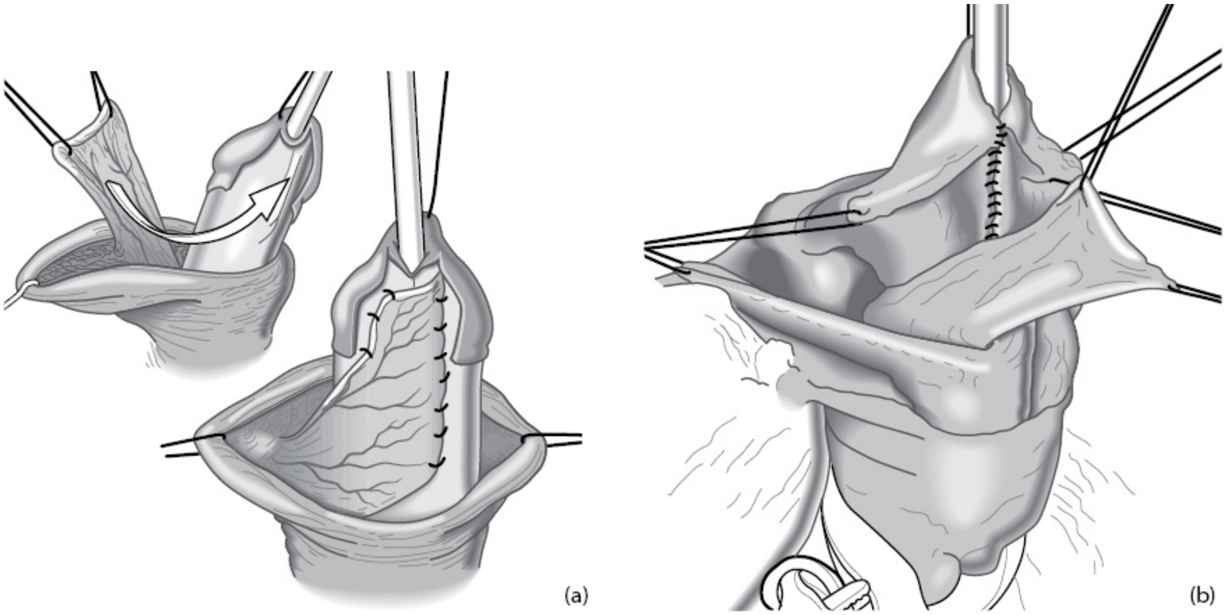
An important surgical principle in neourethra formation is meticulous tissue transfer. In urethroplasty, the tissue source may be adjacent tissue, local tissue flaps, or free grafts of either genital or extragenital tissue origin.

The neourethra may be created by tubularization of the native urethral plate, especially if there is a deep glanular groove. Biologically speaking, the urethral plate contains an epithelial lining with urethral mucosal differentiation and will likely perform best as neourethra, provided that tubularization can occur without suture line tension and meatal stenosis. Local tissue flaps employed for urethral reconstruction must be thin, non-hair-bearing, and easy to handle. They are often derived from distal penile shaft skin or prepuce, and they are called “fasciocutaneous flaps” because of their reliance upon the dartos fascia serving as the conduit for vascular supply and drainage. Graft transfer for neourethra reconstruction is mostly used in the first of a two-stage approach.

Neourethral coverage

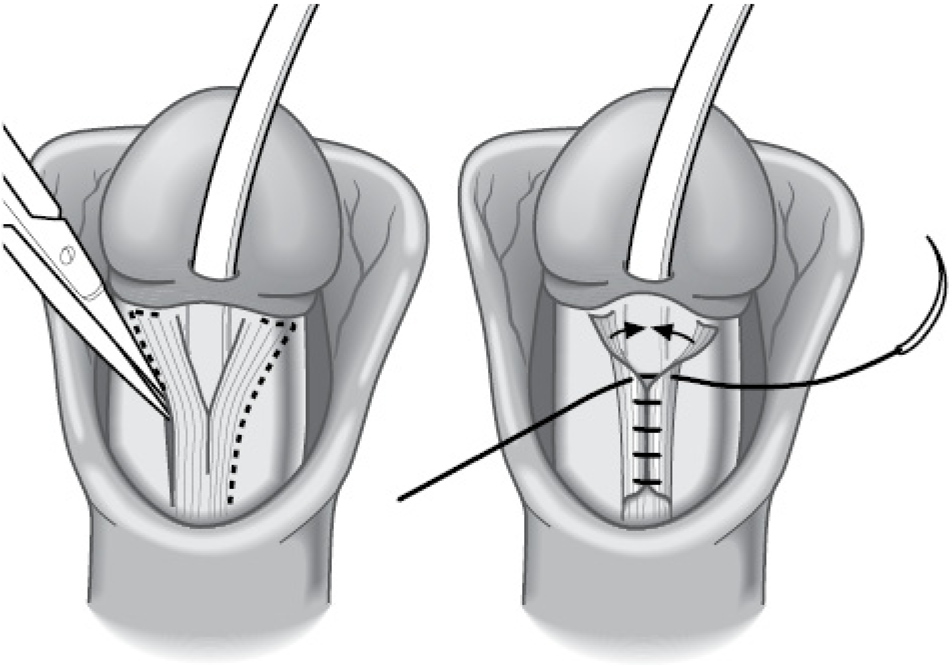
It is critical to establish multiple-layer coverage of the urethroplasty suture line prior to skin closure in order to decrease the risk of urethrocutaneous fistula formation. Various vascularized flaps may be used for this purpose, including dorsal or ventral penile/preputial dartos, tunica vaginalis, and periurethral corpus spongiosum.

The dorsal prepuce is unfolded, and the subcutaneous dartos flap is sharply dissected off the undersurface of the penile skin down to the base and rotated ventrally (**Figure 74.9a**). To prevent penile twisting, the flap may be divided down the middle, preserving the vessels, brought around the shaft to the ventral area, and secured over the neourethra as a double crossover flap (**Figure 74.9b**). Some advocate the use of ventral dartos tissue harvest for neourethra coverage, although this can be difficult in the absence of ample ventral tissue over the dysplastic hypospadiac urethra. For a tunica vaginalis flap, a testicle is delivered and its tunica vaginalis is dissected off the spermatic cord. The flap must be adequately mobilized to avoid ventral tethering and testicular ascent.



74.9a,b

In most cases of hypospadias, the corpus spongiosum becomes flat and often rudimentary distal to the meatus and fans out around the urethral plate. After the urethroplasty, if this tissue layer is robust, it can be mobilized and approximated over the midline as the second layer coverage (**Figure 74.10**). Unlike dartos and tunica vaginalis flaps, the corpus spongiosum used in this way is less likely to cause ventral tethering and penile twisting.



74.10

Meatoplasty and glanuloplasty

All attempts at neourethral reconstruction will be for naught if careful attention is not given to this phase of the hypospadias repair. Glans wings (the lateral flaps of glans tissue that are brought around and approximated over the middle) must be loose with minimal tension, with enough space to accommodate both the neourethra and the coverage flaps. If the glanuloplasty is performed with tension, it can lead to either a troublesome meatal stenosis or breakdown of the distal repair, resulting in dehiscence and recurrence of hypospadias or urethrocutaneous fistula, most often at the subcoronal location. In all techniques of hypospadias repair, the final meatal caliber must be ample (8 Fr for infants and at least 10 Fr for children older than 3 years) with a generous spatulation. It is best to avoid a circumferential suture line around the meatus, which predisposes to meatal stenosis.

INTRAOPERATIVE DECISION-MAKING: USING THE RIGHT TISSUES AND CHOOSING THE RIGHT PROCEDURE

Decision-making begins with assessment of meatal location, penile size, curvature, and the quality of ventral skin over the native urethra. In most patients, hypospadias is repairable using well-vascularized local tissues. In mild glanular hypospadias, simple meatal advancement and glanuloplasty (MAGPI) may be all that is required. In other situations, consideration is given to neourethral reconstruction using either flap technique or urethral plate tubularization. If the urethral plate is healthy with decent width and vascularity, it may be tubularized, either with or without the help of vertical midline incision technique (tubularized incised plate (TIP)/Snodgrass). If it is too narrow or too shallow for effective, tension-free tubularization, a preputial (onlay urethroplasty) or perimeatal-based proximal skin (Mathieu procedure) fasciocutaneous flap is approximated onto the urethral plate as the ventral portion of the neourethra. In some situations, the actual meatal location is distal, but the quality of distal urethra leading up to the meatus is thin and dysplastic, along with near absent penile skin ventrally. This type of hypospadias should be treated as a more severe proximal defect by opening this dysplastic urethra ventrally back to more supple urethral tissue.

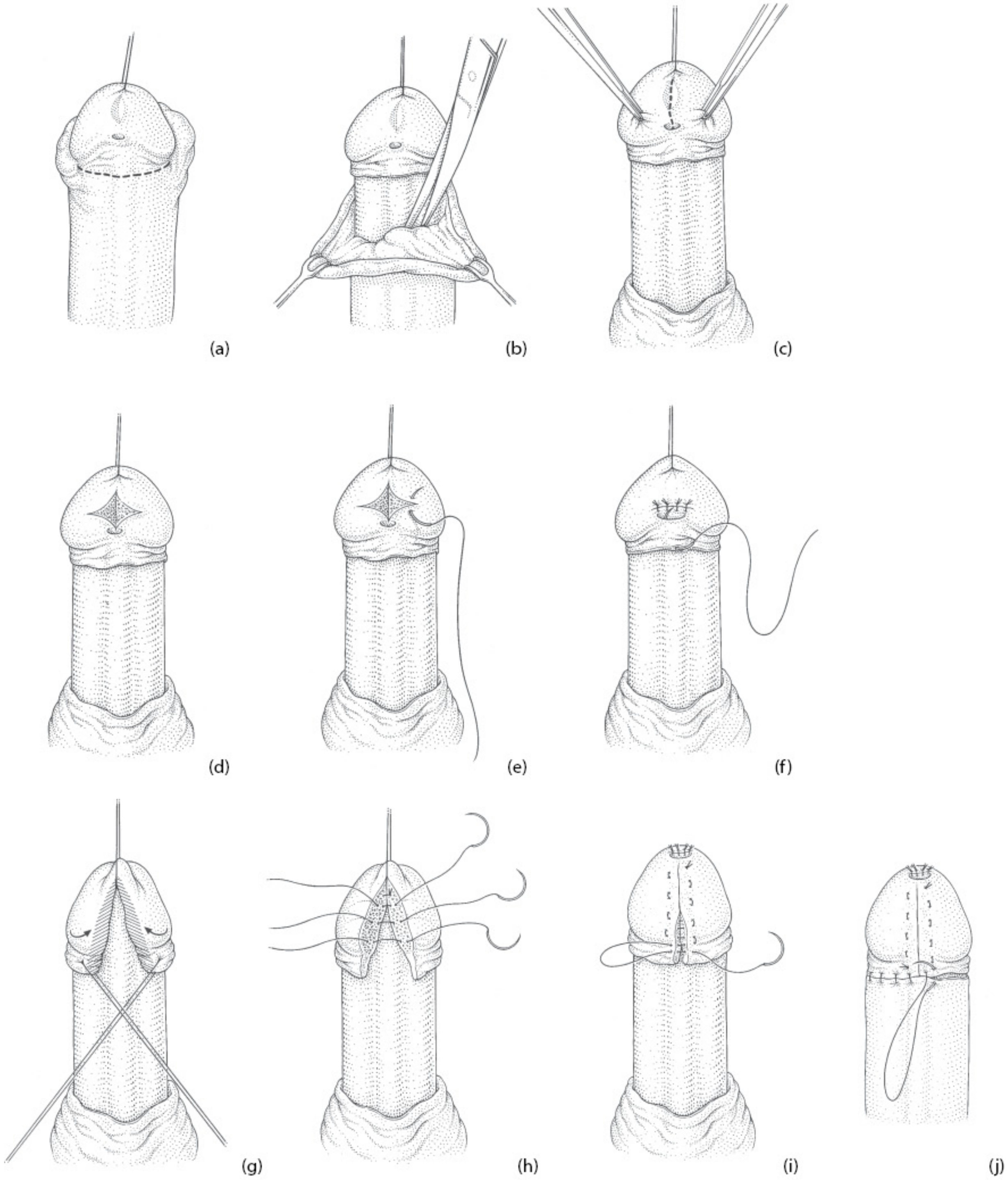
The choice of urethroplasty technique may depend upon the degree of chordee. The penile shaft is degloved after making a subcoronal incision, preserving the urethral plate of 8–10 mm width, except for severe hypospadias such as scrotal and perineal variety in which urethral plate preservation is unlikely from the outset. The penis is evaluated for curvature with an artificial erection test. If the curvature is moderate or severe (greater than 30 degrees), along with short phallic length, correcting chordee by aggressive dorsal corporal plication may not be the best option, since it may lead to shortening of the phallus. In this situation, consideration must be given to division of the urethral plate, excision of ventral fibrous chordee tissues, and use of single or multiple relaxing transverse incisions of the tunica albuginea with or without placement of a corporal patch. The neourethra may then be created later using transposed dorsal preputial skin (staged operation), or immediately with island pedicle flap (transverse island tube urethroplasty), or free graft (oral mucosa graft urethroplasty).

SPECIFIC OPERATIONS

There is no single, universally applicable technique of hypospadias repair. A surgeon attempting an effective hypospadias reconstruction must be familiar with several different techniques, understanding their pros and cons, along with their indications for specific anatomic configurations. It is unwise to rigidly apply one or two "favorite" techniques to all situations. Instead, a given anatomic situation must dictate the procedure to be employed, and the surgeon must maintain flexibility and versatility. It is not possible within the scope of this chapter to discuss all reported techniques of hypospadias repair. The authors will present several of their own preferred techniques.

Meatal advancement and glanuloplasty

For the glanular and some of the coronal hypospadias without significant chordee, MAGPI is a simple, yet elegant, procedure with an excellent functional and cosmetic outcome (**Figure 74.11a–j**). In many cases of distal hypospadias, there is a transverse glanular tissue ridge that separates the true meatus from a distal blind-ending groove. In the first step of MAGPI, this tissue ridge is incised deeply in a vertical direction, creating a diamond-shaped defect, which is then closed in a transverse direction (the Heineke–Mikulicz principle). This maneuver widens, advances, and flattens the urethral meatus. A circumferential incision is then made in the subcoronal region, and the penis is degloved. After addressing any chordee, the glanuloplasty is performed. After ventrolateral de-epithelialization and mobilization of the glans tissue proximal to the urethral meatus, glanuloplasty is performed in two layers, while gently retracting the lower lip of the urethral meatus upward. By avoiding a urethroplasty suture line, there is little to no risk of urethrocutaneous fistula formation, but a meatal retraction back to the hypospadiac position may occur postoperatively if the glanuloplasty is done under tension without adequate mobilization of the glanular wings.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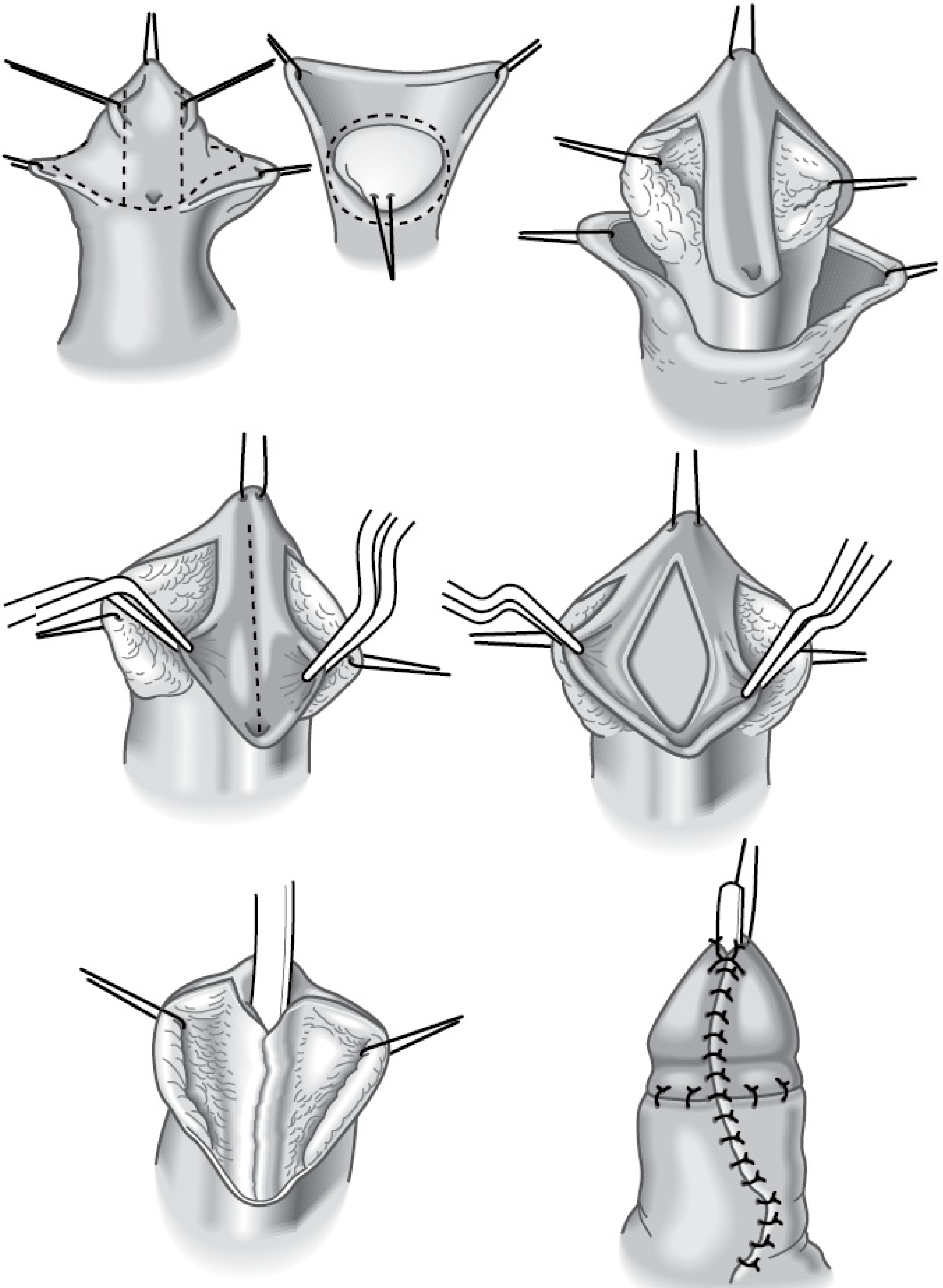
74.11a–j

Tubularized incised plate urethroplasty (TIP or Snodgrass) repair

Thiersch and Duplay were the first to describe the technique of urethral plate tubularization in repairing hypospadias. Additional modifications were described by King and Zaontz (GAP procedure). These techniques all result in excellent cosmetic and functional outcome in patients with hypospadiac penis associated with a deep glanular groove. However, urethral tubularization techniques were not deemed suitable for penis associated with a flat urethral plate, with the fear that there would be an excessive tension of the neourethral suture line along with the risk of meatal stenosis. Snodgrass combined the vertical distal urethral plate incision technique to relax its tension with the Thiersch–Duplay tubularization technique, to propose the TIP repair technique (**Figure 74.12**). Many reports from various institutions have reported excellent outcomes with this procedure. Unlike the traditional urethral plate tubularization techniques, TIP repair is versatile for many different anatomical presentations, including more proximal and previously failed cases.



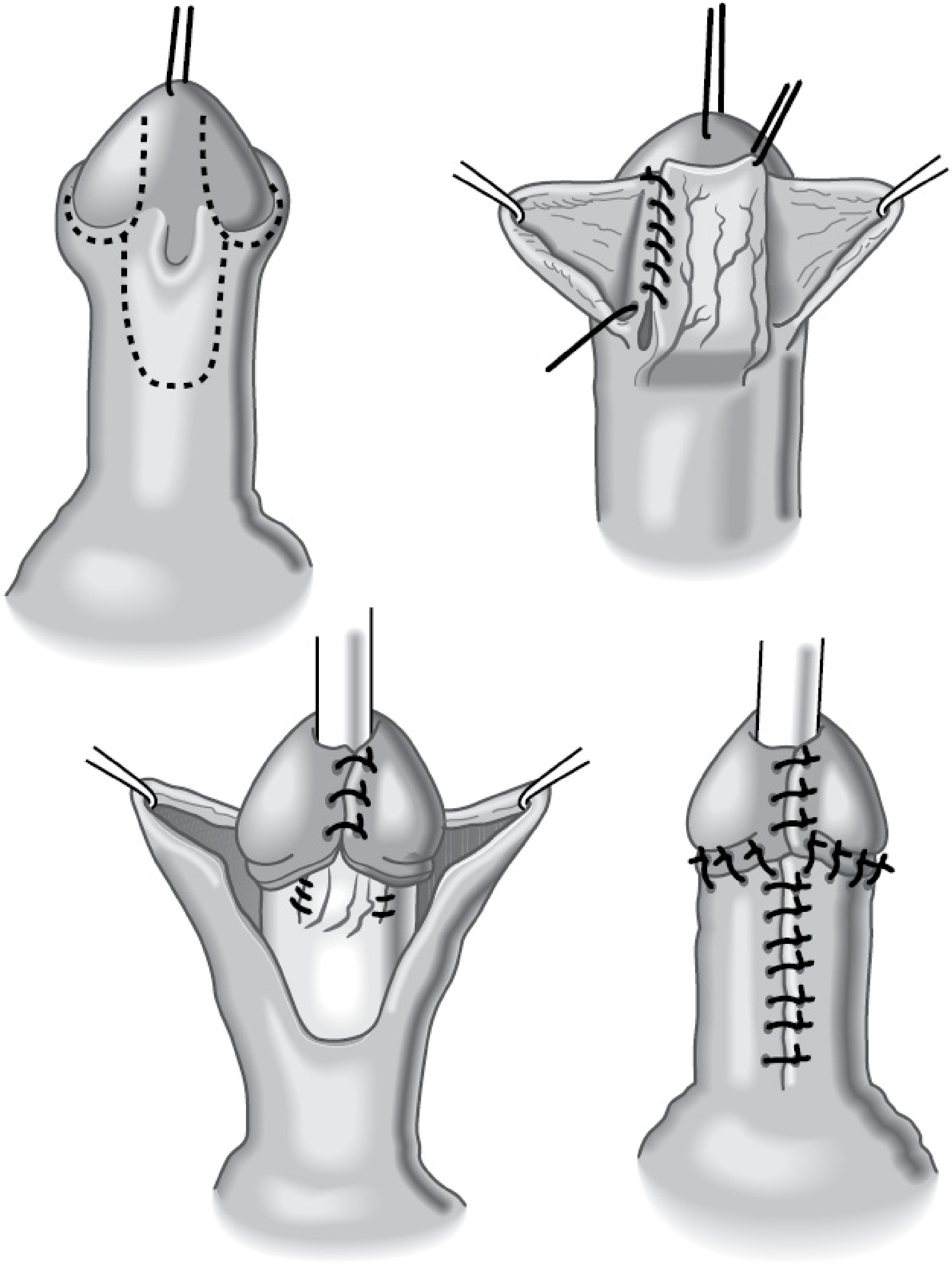
74.12

The urethral plate is marked out 8–10 mm wide distal to the urethral meatus, and incisions are made along the lateral borders of the urethral plate. The distal limit of this incision must be carefully planned, so that at the end of neourethral tubularization, one does not end up with a circumferentially sutured urethral meatus, which may contract and stenose. An incision is made subcoronally and is completed ventrally proximal to the urethral meatus. The penis is carefully degloved down to the base. Ventrally, the skin can be quite thin and adherent to the urethra, and a sharp iris scissor dissection using carefully placed skin hooks for upward counter-traction provides an optimal visualization of the surgical dissection planes to avoid buttonholing into a thin native urethra. After addressing any chordee, glanular wings are adequately developed lateral to the urethral plate and sometimes requiring aggressive dissection over the tips of the corporal bodies to ensure the glanuloplasty can be performed without tension. The urethral plate is gently wrapped around an 8 Fr (10 Fr in older children) tube to check for any areas of tension. While providing a symmetrical traction and countertraction, a deep midline, full-thickness vertical incision is made with a knife in the urethral plate. The neourethra is then reconstructed using multiple interrupted or running sutures. A second layer coverage is then sought with either mobilized dartos, tunica vaginalis, or fanned-out Y-shaped distal spongiosal tissues. Skin coverage of the phallic shaft and glanuloplasty complete the procedure.

Perimeatal-based flap urethroplasty (Mathieu)

Among the more commonly used local flap techniques for coronal and subcoronal hypospadias is the perimeatal based flap technique of Mathieu (**Figure 74.13**). This requires ample penile ventral skin proximal to the hypospadiac meatus. It is begun by measuring the length of the defect from the urethral meatus to the glans. An equal distance flap is then measured from the meatus toward the base of the penis on the proximal shaft skin.

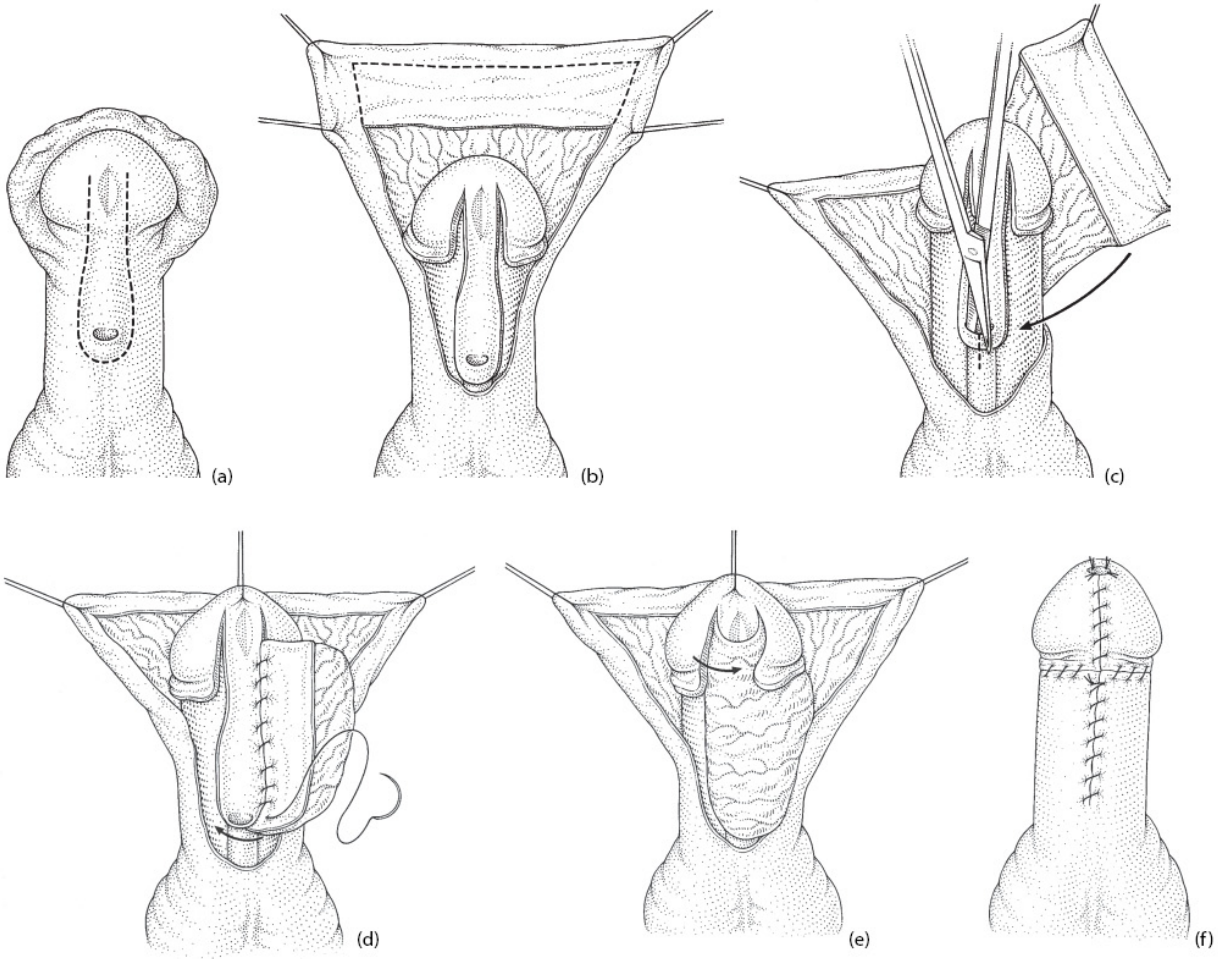
The urethral plate and the matching proximal shaft skin flap are incised to be approximately 7–8 mm wide, and the penis is degloved after subcoronal circumferential incision. Glanular wings are developed deeply in order to perform a tension-free glanuloplasty. If the urethral plate is very flat distally, or if the distal limit of urethral plate is too ventral, the Barcat balanic groove technique may be employed. In this modification, the urethral plate is dissected off the glans tissue, the dorsal glans tissue in the midline is incised, and the urethral plate is advanced further posteriorly to achieve a more distal neourethral opening. After correcting the chordee, the premeasured proximal shaft skin flap is mobilized carefully and transposed upward toward the urethral plate. This flap is folded over the urethral meatus, and both edges are approximated with precisely placed fine absorbable sutures. After maturing a wide caliber meatus, a second layer coverage is performed as described previously.Glanuloplasty and skin coverage complete the procedure.



74.13

Preputial island pedicle flap urethroplasty

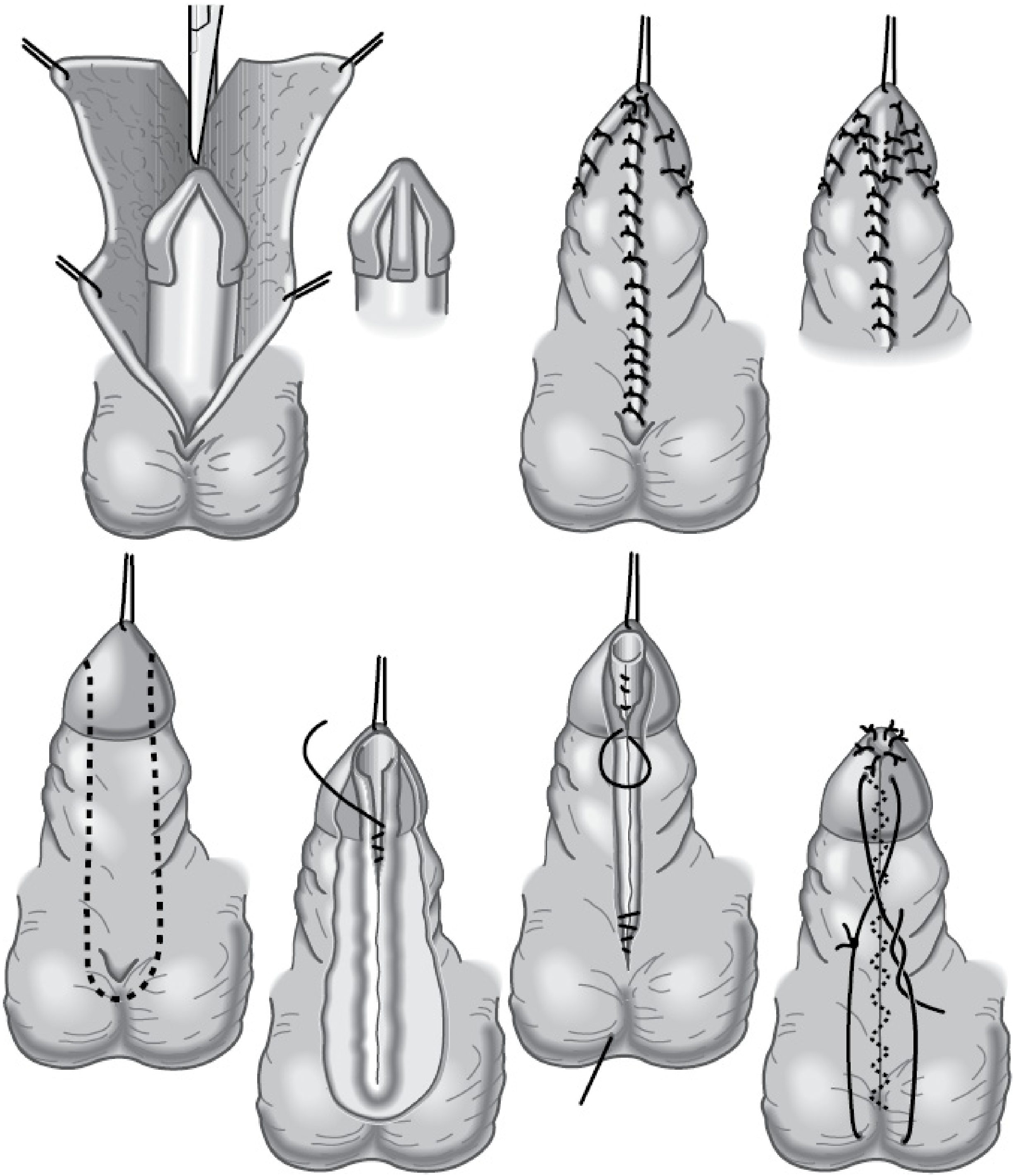
Duckett, Asopa, and Standoli described preputial onlay urethroplasties nearly simultaneously, although the Duckett variant became the popular choice (**Figure 74.14a–f**). With the advent of the TIP repair and increased use of staged repairs, this procedure is used very selectively in those with a more proximal hypospadiac meatus, narrow urethral plate, and minimal chordee. The penis is prepared by means of degloving, and orthoplasty is performed as described above, preserving the urethral plate similar to the TIP repair. The hood of foreskin is laid out and a rectangular portion is marked on the inner surface with a marking pen to outline the graft for the intended neourethra. A rectangular flap is developed using sharp knife and iris scissor dissections, leaving it attached to a broad dartos vascular pedicle. This island pedicle flap must be mobilized adequately down to the penile base in order to swing it away from the remainder of the foreskin and bring it around the penile shaft ventrally without twisting the shaft. The native urethral meatus is prepared to assure that it is vascular, ample, and spatulated. The island flap is then sutured onto the urethral plate using fine absorbable sutures (onlay technique). The glanuloplasty is performed as previously described by creating lateral glanular wings. A critical next step is to provide a supporting vascular tissue to cover the neourethra before skin closure using either the remaining penile dartos or tunica vaginalis. Lastly, ventral skin approximation is accomplished using the dorsal remnants of prepuce.



74.14a–f

Staged repair

Staged repair was once preferred for most instances of hypospadias. Accumulated expertise in hypospadiology relegated staged repairs to the more difficult instances of hypospadias, and some even favored complex single repairs for all instances of hypospadias. Pendulums, however, do swing, and many surgeons prefer to manage patients with complex hypospadias via several dependable steps (**Figure 74.15**). The principles of staged repair are to correct any chordee and other scrotal anomalies (such as penoscrotal transposition) during the first repair, followed by subsequent urethroplasty at another setting. Some patients present with extreme chordee (>90 degrees) along with severe hypospadias. In this scenario, even after straightening the penis perfectly at the time of surgery as proven by an artificial erection test, some recurrence of chordee becomes evident after a number of months. Thus, for the first stage, an initial orthoplasty is performed along with ventral resurfacing with dorsal preputial skin. This resurfacing is accomplished by making a midline dorsal incision of the unfurled inner and outer prepuce. These “Byar’s flaps” are rotated dorsally and the inner preputial skin is fashioned to the apex of the glans wings and approximated in the midline of the degloved ventral surface. At least 6 months later, the patient is brought back for the second stage. Here the residual chordee (which is invariably much less than at the initial procedure) can be corrected definitively, and the neourethra can then be fashioned either by tubularization of the previously transposed preputial tissue or by free graft.

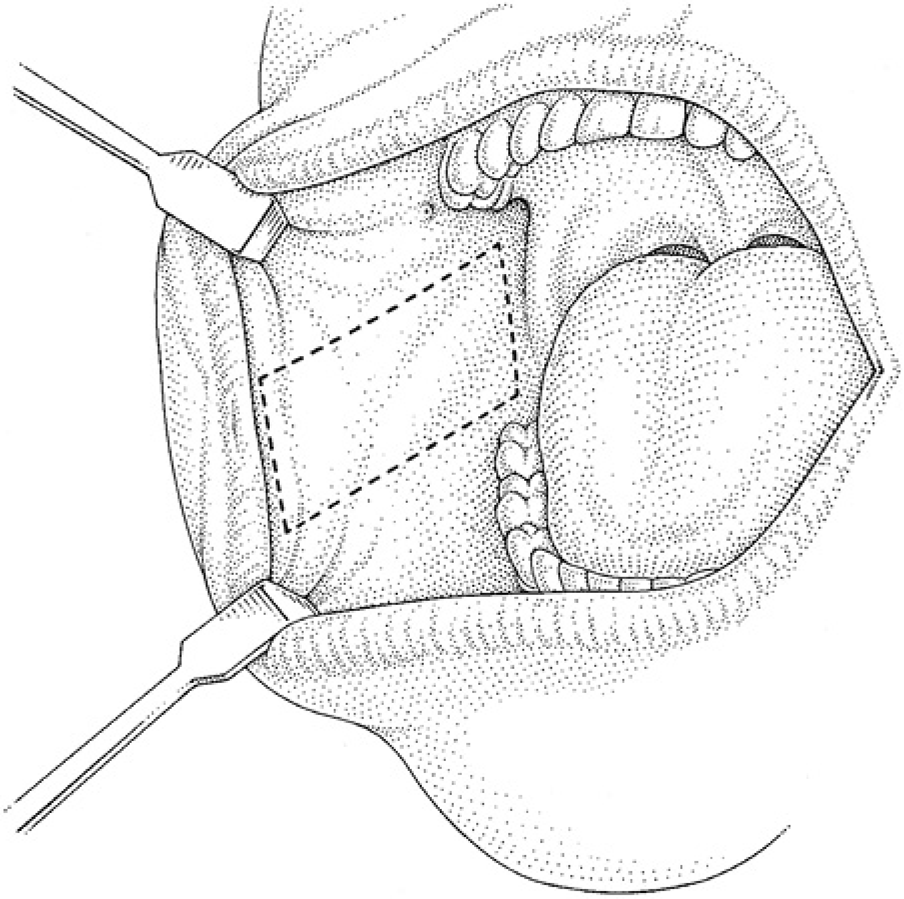


74.15

Oral mucosa graft urethroplasty

Humby reported this technique in 1941 in a single case, but somehow the best hypospadiologists remained ignorant of this innovative technique for more than 40 years until it was rediscovered and resurrected in Italy and France. The next set of papers appeared in the early 1990s, and the use of oral mucosa increased steadily over the next decade in the reconstruction of hypospadias and other urethral problems. This procedure was first applied for hypospadiac "cripples" who had failed multiple operations and were left with no viable local penile tissues for adequate repair, but its use has now expanded satisfactorily for single and staged repairs depending on the anatomic situation.

The oral mucosa graft may be obtained from either the inner lining of the lower lip or inner cheek, and sometimes a contiguous combination of both is useful when a longer graft is needed (**Figure 74.16**). An 8–10 cm long graft can be achieved with this technique. In case of shorter grafts, inner cheek is preferable, and during the harvesting, one must be careful to avoid injury to the parotid duct (Stensen's duct), which is opposite the second upper molar in most patients. The graft harvest site is marked out with a marking pen, retracting the mouth with a combination of retractors and fine traction sutures. Subcutaneous injection of dilute (1 : 200 000) epinephrine solution is useful for purposes of hemostasis and facilitating the dissection of the oral mucosa. A sharp dissection is performed using knife and iris scissors, leaving muscle bundles in the mouth. Bleeders in the graft bed are managed with a combination of direct pressure, pinpoint coagulation, or fine suture ligatures. The harvest site may be closed with fine absorbable sutures (such as 4/0 chromic), or it may be left open. No clear-cut advantage has been demonstrated for either strategy, and postoperative morbidity in terms of pain and dietary issues has been negligible. Most patients can resume a normal diet in 24–36 hours. The underside of the graft is then carefully trimmed to remove any extraneous adipose and muscle tissue. During the entire oral procedure, the initial operative field over the penis and the surgical instruments are segregated before the mouth is prepared and draped.



74.16

After completing the graft harvest and preparation, the surgeons reglove and gown and return to the original operative field and instruments. The graft is then employed for creating neourethra via either ventral onlay (if an adequate urethral plate is present) or dorsal onlay on the exposed ventral penile tissue using a staged technique for a revision repair. Prior to dorsal onlay, the graft can be fenestrated with small full-thickness incisions to allow drainage and therefore decrease graft separation from the underlying wound bed if a hematoma or seroma were to form. For dorsal onlay staged repairs, a bolster is often secured over the mucosal surface to increase graft adherence. Patient's activity must be restricted for 24–48 hours to encourage vascular ingrowth into the graft.

POSTOPERATIVE CARE

Urethral catheter

Intuitively, a newly reconstructed urethra with a long suture line is likely to be better protected during healing with the use of diverting urethral catheter. It is routine among many surgeons to leave an indwelling urethral catheter postoperatively for 7–10 days. However, recent studies have not proven the benefit of urethral catheter. In a multicenter report, excellent results were obtained in 96.7% of over 300 patients undergoing mild to moderate hypospadias repair with the Mathieu technique, and the complication rate was not affected by the catheterization status. Others have speculated that the routine use of urethral catheter may at times increase the chance of complications. Based on these observations, a routine use of urethral catheter may not offer any significant advantage in mild to moderate cases of hypospadias with a straightforward neourethra reconstruction. If the authors were to use a catheter, a soft Silastic tube of 8–10 Fr caliber is preferred. The urine is drained directly into a diaper without a drainage bag. Sometimes the double diaper technique is utilized in infants, in which the catheter is brought through a ventral hole in the inner diaper and is allowed to continuously drain into the outer diaper. With this set-up, fecal material is kept away from the catheter opening, and the double diaper provides a secure additional padding over the genitalia without the worry of a drainage bag pulling on the reconstructed urethra.

Dressing

An ideal penile dressing after hypospadias repair should be non-adherent, absorbent, and compressive, while being soft and elastic enough to accommodate postoperative swelling. Several variations have been reported in the literature. A careful application of secure penile dressing can prevent postoperative complications, such as hematoma and edema, and may additionally reduce parental anxiety. It is critical to keep the reconstructed meatus moist and free of dried secretions by generously applying a petroleum-based ointment for several weeks.

COMPLICATIONS

Bleeding

In most hypospadias repairs, electrocautery must be used very sparsely, applying precise pinpoint cauterizations. Bleeding from the exposed spongiosal tissues, from the glans, corpus spongiosum, or corpus cavernosum, should not be managed with aggressive cauterization, not only because of ineffectiveness in stopping the bleeding but also because of excessive tissue ischemia and injury following such maneuvers. Dilute epinephrine solution, direct pressure, or fine suture ligatures are appropriate for most intraoperative bleeding. Additionally, the bleeding is often coming from beneath the glans wings and can be difficult to identify without deconstructing the repair, which is not advisable. Pressure dressings are useful for hypospadias repairs with the caveat that they should not be too tight to cause ischemia. Occasionally, a patient will return to the emergency room a day or so after hypospadias repair because of unanticipated bleeding, and in these instances, we will simply reapply a pressure dressing. It may be necessary on rare occasions to return to the operating room to evacuate a clot and control a bleeding source. Late bleeding, more than a week postoperatively, is unusual and may occur from trauma. Large expanding hematomas are best evacuated under anesthesia, with control of active bleeding sites and reapplication of a pressure dressing.

Meatal stenosis

A wide distal anastomosis without a circumferential suture line, while preserving good vascularity to all the involved tissues, should minimize subsequent meatal stenosis. In the initial few postoperative months, it is critical to keep the distal meatus continuously moist with petroleum-based ointment. When we see the patients back in clinic, we have a low threshold for passing a small caliber feeding tube to assure patency. When meatal stenosis is suspected, we will often ask the families or the patients to pass a tube on a regular basis to keep it open. Topical vitamin E or corticosteroids may help soften an incipient stricture. A late stricture may require an anesthetic for aggressive dilation or urethrotomy. It is important to distinguish true meatal stenosis from distal urethral stenosis, which would require more aggressive management and even redo urethroplasty.

Urethrocutaneous fistula

A very distal fistula in the glans or corona can be corrected by simply incising the intervening glanular tissue to create a more ample meatus, even if it becomes slightly hypospadiac, as long as there is no chordee. More proximal fistulas are corrected by generous incision to expose the defect and multiple layers of closure, usually using some type of de-epithelialized skin edges to create vascular coverage layers. Some fistulas represent only the tip of an iceberg, as it were, and are indicative of unhealthy tissue over a portion of the neourethra or distal urethral stricture. In these instances, one must consider doing a complete revision of neourethra with alternative tissue sources, such as the oral mucosa. Not appreciating the poor quality of neourethra as the true cause of fistulas will likely lead to their recurrence. In many instances, the *sine qua non* for a successful fistula repair is: “Make it a big operation to fix a little hole.”

Infection

True skin infections with cellulitis are uncommon after hypospadias repair. When they are suspected, however, an aggressive antibiotic treatment with broadened coverage should be initiated, and the child brought back into hospital for several days of IV antibiotic coverage. When the children are sent home with a catheter in place, they are routinely given a low-dose prophylactic antibiotic coverage daily to minimize colonization and reduce the risk of UTI until the catheter is removed. However, the benefit of this is questioned in the literature.

Urethral diverticulum

This complication occurs as the result of either distal urethral stenosis and/or insufficient ventral tissue coverage of the neourethra. A small diverticulum may be left alone if a patient is willing to support the sacculed area manually while voiding; however, more substantial urethral diverticuli are best managed surgically, first by ensuring no distal stenosis and second by exposing the diverticulum, removing excess tissue, and reclosing the urethra. The critical step is finding adequate tissue (either dartos or tunica vaginalis) to support the area of the resected diverticulum before closing the skin. Again, if a significant portion of urethra is unhealthy or ischemic, it is better to perform a revision urethroplasty using an oral mucosa graft.

Stricture

Urethral stricture may occur at the meatus (meatal stenosis), in the glanular portion of the urethra, or at a proximal anastomotic site. Strictures within the middle portion of the neourethra are less common. Early strictures are best managed by gentle dilation and intermittent catheterization. More severe strictures, particularly along the course of the urethra, may be corrected by a dilation technique or internal urethrotomy using either endoscopic knife or laser. Generally speaking, if the strictured segment is long and is surrounded by dense periurethral fibrosis, the above-mentioned minimally invasive techniques are ineffective, and the stricture is prone to recur. It is advised that, in these situations, one should consider a revision urethroplasty using oral mucosa.

Reoperative hypospadias repair

Some patients present with a difficult constellation of failed hypospadias repair, residual chordee, recurrent stricture, fistula, or other functional residua after multiple attempts at repair. These patients have been called “hypospadiac cripples.” The fundamental principle of management is thorough assessment of the relevant symptoms and anatomic features of the patient. Most often, this is best done by examination under anesthesia with cystourethroscopy. The first principle is to properly inform the patient and family about the problems and likely etiologies, plan for correction, and potential complications. Establishing realistic expectations is critical. The second principle is to recognize any related anatomic features, such as penoscrotal transposition and chordee, as well as any available viable tissues, not only for neourethra formation and penile skin coverage but also vascular supporting tissues for covering the neourethra. At times, planning a staged approach, correcting chordee, and debriding unhealthy local tissues before proceeding with neourethra reconstruction, may be prudent. Third, the neourethra must be reconstructed from robust, mucosa-lined tissues with an excellent potential for neovascularity (most likely this will be oral mucosa). Stubbornly insisting on using the remnant of fibrotic preputial and penile skin will lead to an inevitable failure and poor long-term outcome. Finally, a period of postoperative catheterization, early reassessment with calibration of the neourethra, and even a confirmation endoscopic examination when appropriate, will be essential.

FURTHER READING

Baskin LS, Duckett JW. Dorsal tunica albuginea plication for hypospadias curvature. *J Urol* 1994; 151(6): 1668–71.

Chua M, Welsh C, Amir B, et al. Non-stented versus stented urethroplasty for distal hypospadias repair: A systematic review and meta-analysis. *J Pediatr Urol* 2018; 14(3): 212–19.

Duckett JW Jr. Transverse preputial island flap technique for repair of severe hypospadias. *Urol Clin North Am* 1980; 7(2): 423–30.

Duckett JW, Coplen D, Ewalt D, Baskin LS. Buccal mucosal urethral replacement. *J Urol* 1995; 153(5): 1660–3.

Gong EM, Cheng EY. Current challenges with proximal hypospadias: We have a long way to go. *J Pediatr Urol* 2017; 13(5): 457–67.

Park JM, Faerber GJ, Bloom DA. Long-term outcome evaluation of patients undergoing the meatal advancement and glanuloplasty procedure. *J Urol* 1995; 153(5): 1655–6.

Retik AB, Borer JG. Primary and reoperative hypospadias repair with the Snodgrass technique. *World J Urol* 1998; 16(3): 186–91.

Snodgrass W. Tubularized, incised plate urethroplasty for distal hypospadias. *J Urol* 1994; 151(2): 464–5.

75

Orchidopexy

JOHN M. HUTSON

PRINCIPLES AND JUSTIFICATION

An undescended testis is a common abnormality, affecting 3–5% of boys. In preterm infants, the incidence may be as high as 20%. Many of these not in the scrotum at birth descend by 12 weeks after birth, so that by 3 months of age the incidence of congenital undescended testes is approximately 1–2%. Some testes that descend late into the scrotum may reascend later in childhood, producing an acquired variant of maldescent, known as the ascending testis.

Embryology

The urogenital ridge forms on the posterior abdominal wall and contains the mesonephros, its draining duct, and the developing gonads. Primitive germ cells migrate from the yolk sac in the sixth week of gestation, as the gonad in the male develops into a testis. By 7–8 weeks, the testis is producing hormones that control its subsequent descent.

Descent of the testes

Descent of the testes occurs in two morphologically and hormonally distinct phases. The key structure in controlling the process is the gubernaculum, which is the embryonic ligament anchoring the testis and urogenital ridge to the inguinal region. The gubernaculum enlarges in the first phase to anchor the testis near the inguinal region as the embryo enlarges between 10 and 15 weeks of gestation. In the second phase, which occurs between about 25 and 35 weeks of gestation, the gubernaculum migrates through the inguinal canal, across the pubic region, and into the scrotum. The processus vaginalis (PV) develops as a peritoneal diverticulum within the elongating gubernaculum, creating an intraperitoneal space into which the testis can descend.

The main hormone controlling the first phase is the homolog of insulin and relaxin, known as insulin-like hormone 3 (INSL3). This hormone is produced by Leydig cells, and stimulates the distal gubernaculum, with some augmentation of its effect by anti-Müllerian hormone (AMH), also known as Müllerian inhibiting substance (MIS), and possibly testosterone. In the second phase, testosterone acts partly indirectly via the genitofemoral nerve, which supplies the gubernaculum and scrotum. Calcitonin gene-related peptide (CGRP) has been identified within sensory branches of the genitofemoral nerve and has been postulated to modulate androgenic control of descent. Androgens cause masculinization of the sensory branches of the genitofemoral nerve between 15 and 25 weeks of gestation, and the masculinized nerve then controls the direction of gubernacular migration by the release of CGRP in the periphery.

Some undescended testes in humans may be caused by androgen or INSL3 deficiency, and also from physiologic or anatomic abnormalities of the genitofemoral nerve or its neurotransmitter, CGRP. The common cause of maldescent is failed gubernacular migration, leaving the testes in the groin, or so-called “superficial inguinal pouch,” caused by abnormal anatomic or hormone development. Rarely, undescended testes are caused by recognized anomalies in the hypothalamic–pituitary–gonadal axis or in the secretion or action of androgen, INSL3 and AMH. Recognizable hormonal syndromes, however, are rare causes of cryptorchidism in clinical practice.

Undescended testes

Failed gubernacular migration leads to arrest of the gubernaculum along the normal pathway or aberrant migration to an ectopic location in the perineal, prepubic, or femoral region. Many undescended testes are located in the superficial inguinal pouch, the subcutaneous space just outside and lateral to the external inguinal ring containing the PV (with its contained testis). This is the most common position for an undescended testis and is no longer categorized as ectopic.

There is controversy about whether retractile and "ascending" testes are abnormal, but they really should be viewed as a "grey zone" between normally descended and undescended testes. These testes are located in the scrotum by 12 weeks of age, but later in childhood retract out of the scrotum, and often have an exaggerated cremaster reflex. Many, but not all retractile testes develop into acquired undescended testes, with the testis becoming relatively higher as the child gets older. In most cases, the cause of this is failure of the spermatic cord to elongate normally, while the distance between the inguinal canal and scrotum doubles (from about 5 cm to 10 cm) with growth during the first decade. The cause of acquired undescended testes appears to be failure of the PV to completely disappear after the testis reaches the scrotum.

Surgical intervention is not indicated unless the testis is no longer residing spontaneously in the scrotum. If the testis is near the top of the scrotum, regular follow-up is wise to make sure that it does not ascend out of the scrotum as the boy grows. This often leads to orchidopexy being recommended in 3- to 11-year-old boys.

Indications

Surgery is recommended for three common reasons: Abnormal fertility, a risk of testicular tumors in adult life, and the obvious cosmetic abnormalities. Although many undescended testes have a patent PV, this is an uncommon presentation for an inguinal hernia. If an infant presents with an inguinal hernia, however, orchidopexy with associated herniotomy is performed immediately. Trauma and torsion in undescended testes are more common than when the testis is descended, although these are unlikely indications for surgery.

Germ-cell development in cryptorchidism is relatively normal at birth, but then transformation of neonatal gonocytes into sperm stem cells during “minipuberty” at 3–6 months is abnormal, due to secondary degeneration. Congenital undescended testes leads to too few spermatogonial stem cells, leading to subsequent infertility. This is caused by the testes residing at a higher temperature (35–37 °C) than when in the scrotum (33 °C). In some instances, especially in bilateral cryptorchidism, there may be an intrinsic anomaly of testicular function.

The risk of malignancy in young adults with undescended testes is approximately fivefold higher than in young men with descended testes, and is thought to be caused by abnormal persistence of some neonatal gonocytes. These risk calculations are not based on current practice, however, but on results when children had orchidopexy at approximately 10 years of age. With the current practice of recommending surgery for congenital undescended testes at 6–12 months of age, it is hoped that the cancer risk in the next generation will be lower, although this remains unproven. The risk of malignancy affects not only the unilateral undescended testis but also the contralateral descended testis in some patients. Evidence is now emerging that acquired cryptorchidism has a much lower risk of subsequent malignancy.

Recommended age for orchidopexy

To prevent secondary testicular degeneration with loss of germ cells and increasing risk of malignancy, orchidopexy for congenital cryptorchidism is recommended between 6 and 12 months of age. Although it is not proven for humans, it is clear in animal experiments that early surgery is better than a delayed operation. Operation can be performed at any time between 3–6 months and 1 year of age, depending on the surgeon’s experience. Those with less experience would be wise to delay surgery to the older end of the range, rather than to attempt orchidopexy in a young infant, to avoid testicular atrophy. Magnification should be mandatory for surgery in infants <1 year of age.

PREOPERATIVE PREPARATION

In older boys with possible ascending testes, a course of gonadatrophins occasionally may be appropriate, although this is controversial. In bilateral impalpable testes, chromosome analysis and ultrasonography may be needed, and a human chorionic gonadotrophin (hCG) stimulation test or AMH measurement might be performed to determine whether testicular tissue is present at all. Serum levels of AMH will reflect the presence of Sertoli cells. If hCG stimulation shows the presence of Leydig cells, laparoscopy is indicated at the beginning of the operation to determine the exact site and nature of the intra-abdominal testes.

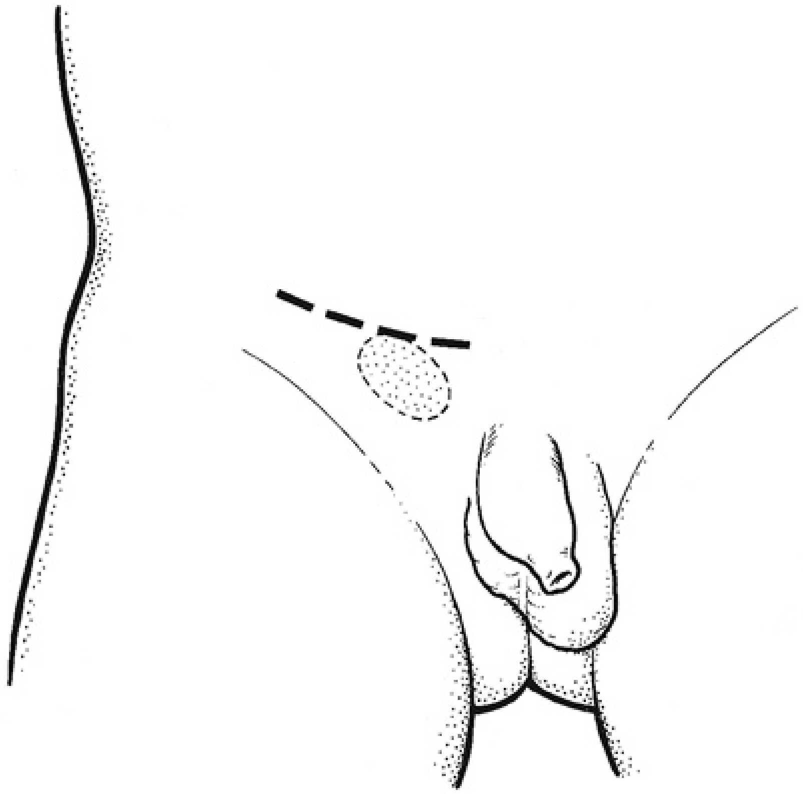
OPERATIONS

Standard orchidopexy

Standard orchidopexy for a palpable testis (which occurs in approximately 80–90% of cases) involves an inguinal incision, full exposure of the inguinal canal, separation of the PV, and mobilization of the testis and spermatic cord. The second part of the operation is the orchidopexy itself, i.e. fixation of the testis in the scrotum.

MOBILIZATION OF THE TESTIS

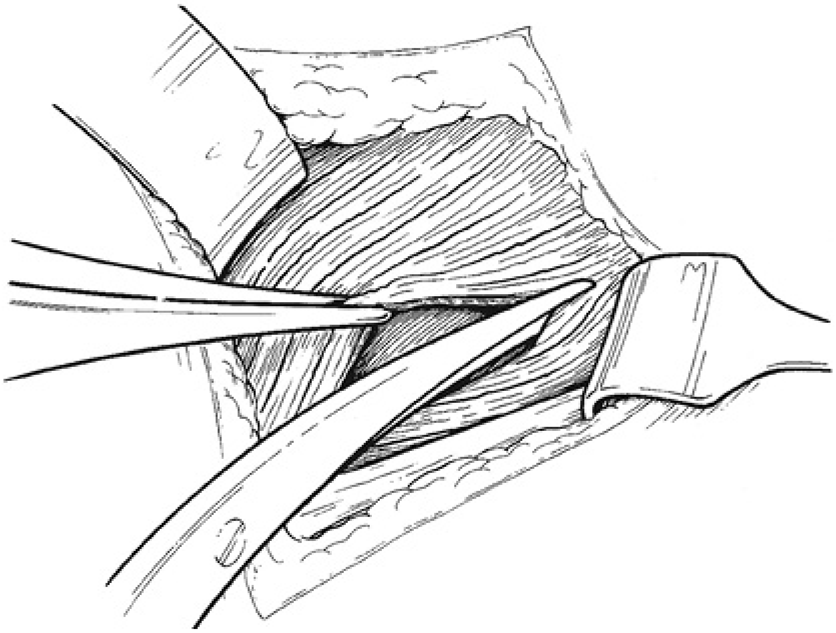
A transverse skin crease incision is made over the inguinal canal. This incision is usually about one finger's breadth above the base of the penis in an infant. The medial end of the incision is level with the pubic tubercle, while the lateral end is at the mid-inguinal point (**Figure 75.1**).



75.1

The incision is deepened through the subcutaneous fatty tissue with scissors or diathermy. The superficial fascia is in two layers: A more superficial fatty layer (of Camper) and a deeper, well-developed fibrous layer known as Scarpa's fascia. The superficial inferior epigastric vein may be seen in the subcutaneous tissue running obliquely across the incision and should be coagulated and divided.

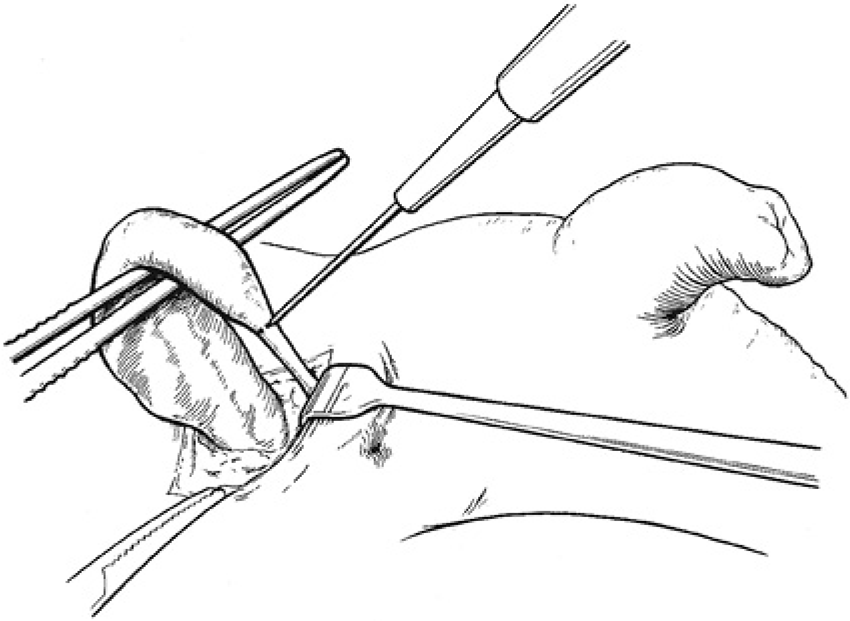
Once Scarpa's fascia has been divided, the external oblique aponeurosis can be distinguished deep to Scarpa's fascia by the oblique orientation of its fibers (**Figure 75.2**). The surface of the external oblique muscle is cleared to expose the lower border of the external oblique muscle where the rolled edge of the inguinal ligament lies and then the site of the external inguinal ring, where the spermatic cord is seen bulging. The inguinal canal is opened with a scalpel incision in the external oblique muscle, with extension with scissors in line with the external ring. The edges of the external oblique muscle are best stabilized with small artery forceps so that they can be identified easily later in the operation.



75.2

The ilioinguinal nerve will run parallel to the incision, just under the external oblique aponeurosis, and should be identified and carefully avoided. Accidental transection of the nerve will produce a sensory deficit in the region of the anterior scrotum. Blunt dissection is used to mobilize the inner layer of the external oblique aponeurosis off the surface of the spermatic cord, and the external ring is opened with scissors if this has not already been done.

The testis and attached spermatic cord are mobilized with blunt dissection and delivered out of the wound (**Figure 75.3**). This should identify the abnormal attachment of the gubernaculum, which often causes dimpling of the skin above and lateral to the neck of the scrotum. The gubernacular attachment is divided carefully with scissors or diathermy, taking care to avoid any structures within the PV, such as the vas deferens, which may extend below the lower pole of the testis. The gubernaculum at this level is usually transparent, with occasional fine vessels and some fat, and is easy to divide without risk to other structures.



75.3

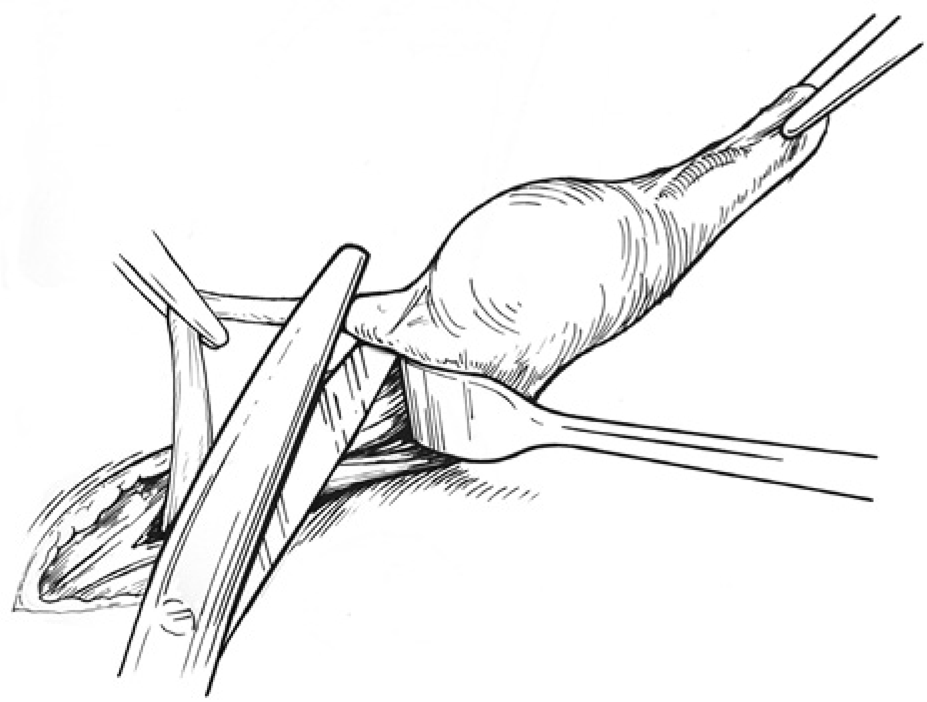
Artery forceps are placed on the gubernacular attachment to the testis and the tunica vaginalis and this is placed on tension. This enables any remaining cremaster fibers surrounding the outside of the spermatic cord to be stripped off with blunt-ended forceps.

DISSECTION OF THE HERNIAL SAC

The PV is commonly widely patent in the undescended testis. Careful separation of the patent PV or obvious hernial sac from the vas deferens and the testicular vessels is an important part of the procedure, as this increases the effective length of the spermatic cord. The hernial sac may be stretched over the index finger while round-ended, non-toothed dissecting forceps gently sweep off the other cord structures, carefully avoiding direct application of the forceps on the vas deferens or vessels. Alternatively, the sac can be held with small artery forceps while the vas deferens and testicular vessels are isolated *en masse* off the sac. It is best to attempt to separate the sac completely without opening it, as an unrecognized tear may extend through the internal ring into the peritoneum, making closure of the hernial sac difficult. It is essential that the testicular vessels, and particularly the vas deferens, are visualized clearly before the sac is divided.

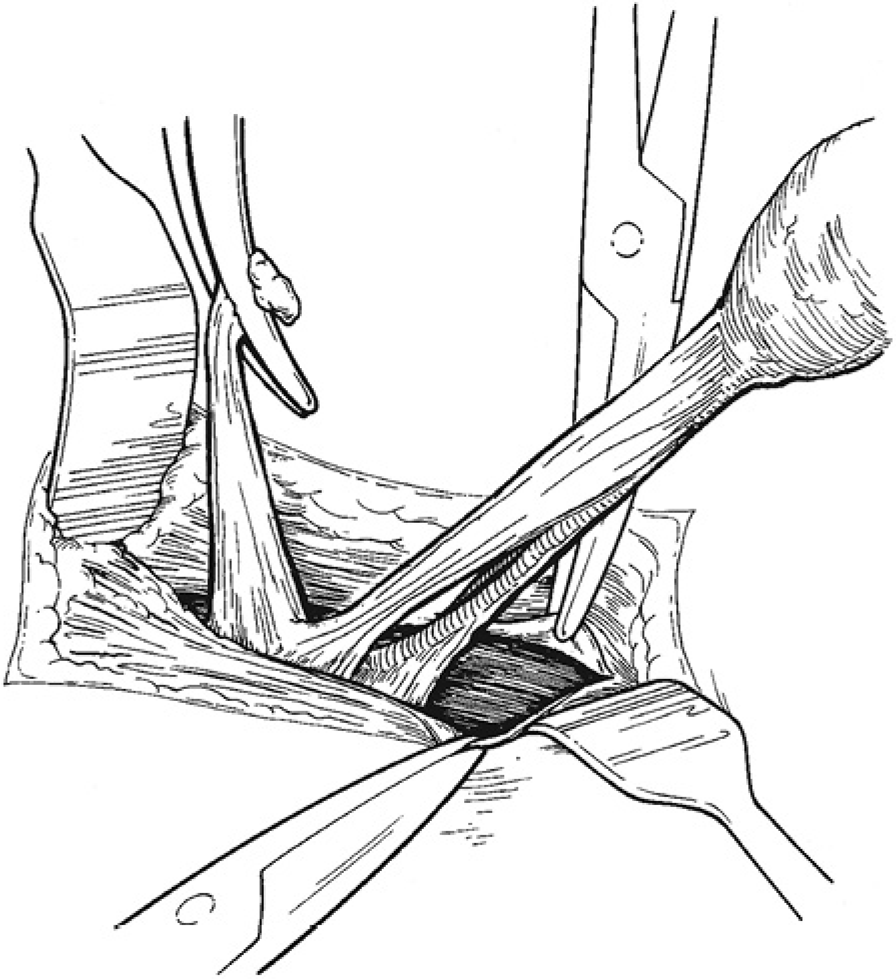
It is important to note that the vas is closely adherent to the posterior surface of the sac before dissection.

With the cord on traction, the testicular vessels and vas deferens are separated from the hernial sac with a small retractor and adequately identified before clamping the hernial sac with artery forceps and dividing it with scissors immediately distal to the artery forceps (**Figure 75.4**).



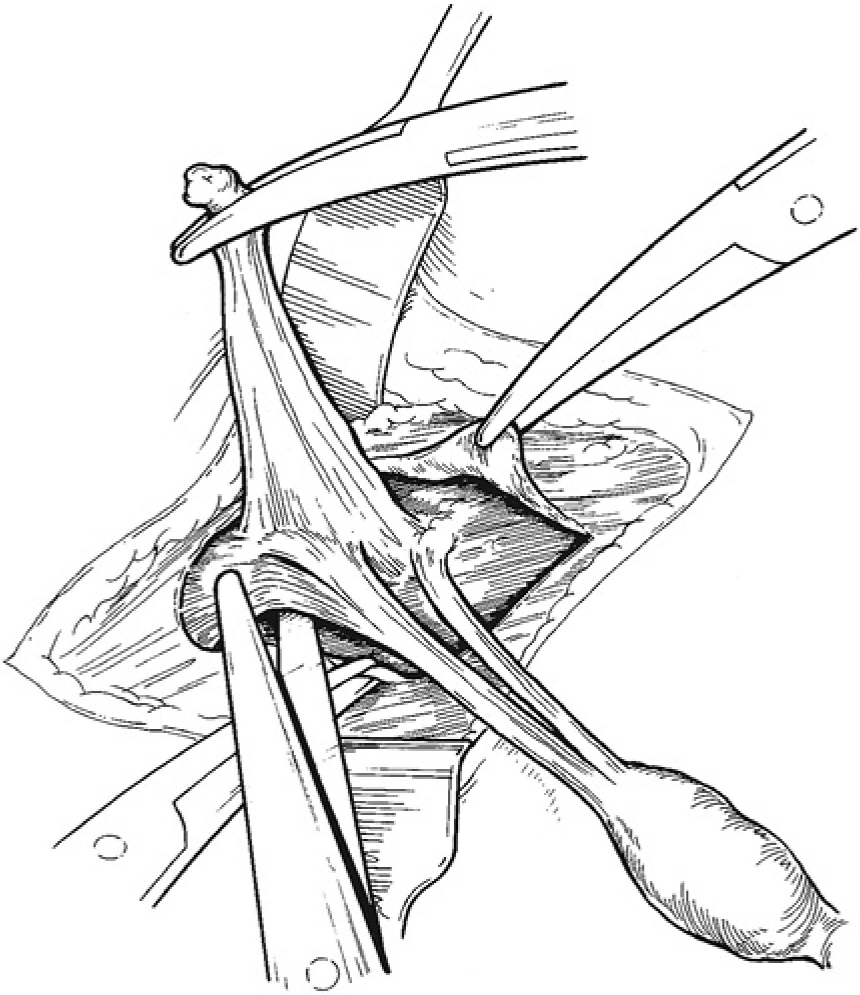
75.4

The divided hernial sac/PV is pulled cranially and the remaining testicular vessels and vas deferens are then separated with blunt dissection from its posterior surface sac right up to the internal ring (**Figure 75.5**). At the junction between the PV and the peritoneum proper, its translucency becomes an opaque, white membrane with triangular widening at the base. At this level, the vas curves medially around the edge of the transversalis fascia, adjacent to the inferior epigastric vessels. By contrast, the testicular vessels pass cranially on the lateral side of the internal ring and disappear into the retroperitoneal space.



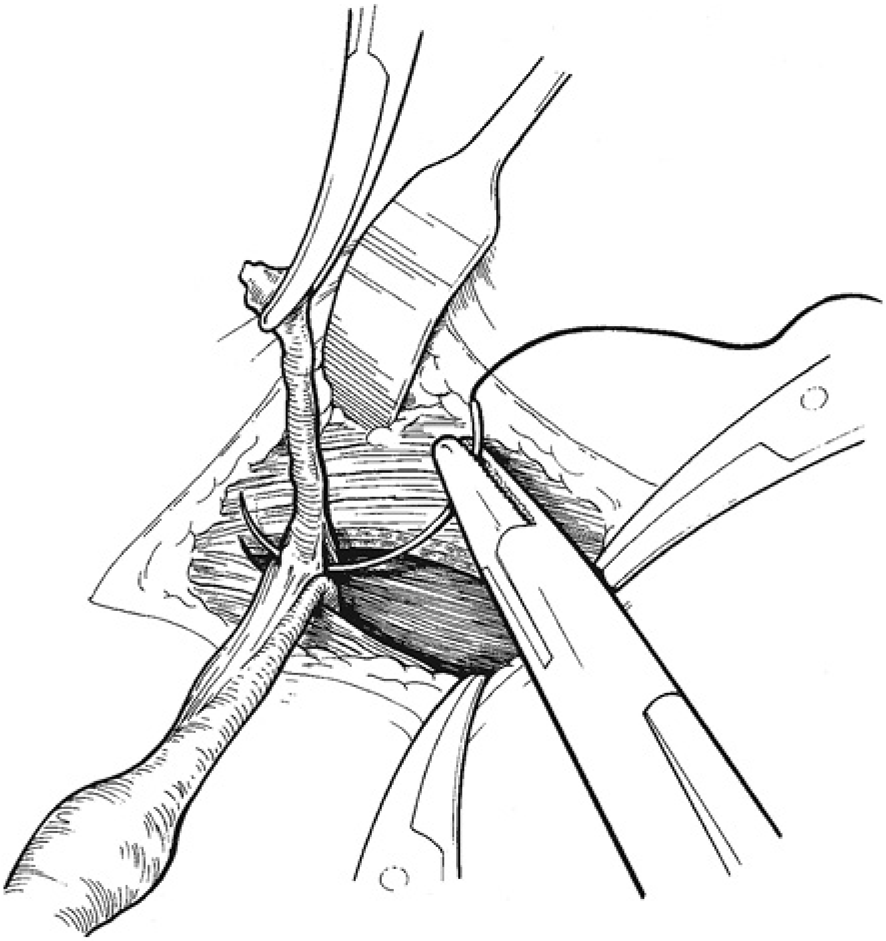
75.5

If the spermatic cord is not long enough to reach the scrotum, the retroperitoneal plane behind the PV above the internal ring is developed, and a Langenbeck's retractor is inserted to pull the peritoneal membrane anteriorly. This shows the testicular vessels passing cranially in the retroperitoneum. The vessels tend to follow a gentle convex curve laterally, and there are a number of lateral fibrous bands (of Ladd) attached to the vessels. These should be divided by sharp or blunt dissection (**Figure 75.6**). Continuous, gentle traction on the testis allows the testicular vessels to be seen and preserved.



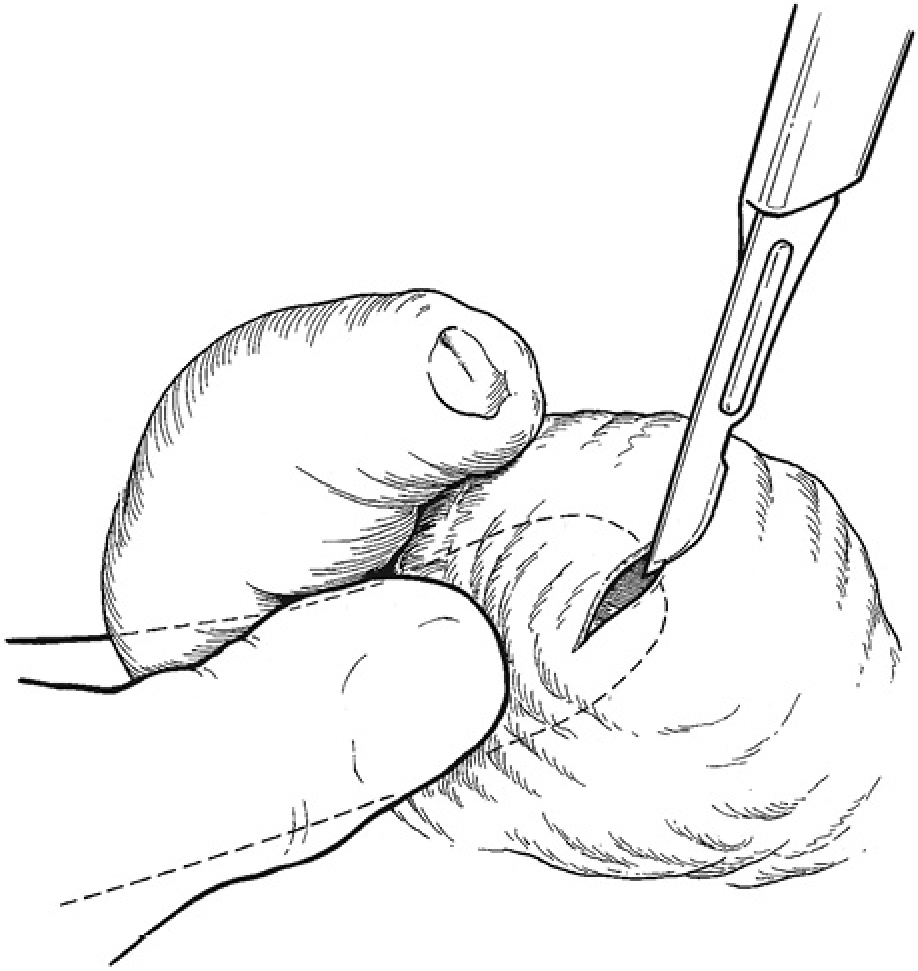
75.6

There should now be adequate length of vas deferens and vessels to allow the testis to reach the scrotum. The PV is now twisted up to the internal ring to make sure that it contains no intraperitoneal contents, and it is transfixed and ligated at this level (**Figure 75.7**). At this point, if the length of the vas deferens is a limiting factor in the position of the testis, the inferior epigastric vessels can be divided to allow the vas deferens to pass around the conjoined tendon rather than the internal inguinal ring. This is known as the *Prentiss maneuver,* and effectively gains an extra 0.5–1 cm. Alternatively, a right-angle forcep can be placed through the internal ring and the back of the inguinal canal opened medial to the preserved inferior epigastric vessels. The testis with its attached vas deferens and vessels can then be brought behind and medial to the inferior epigastric vessels.



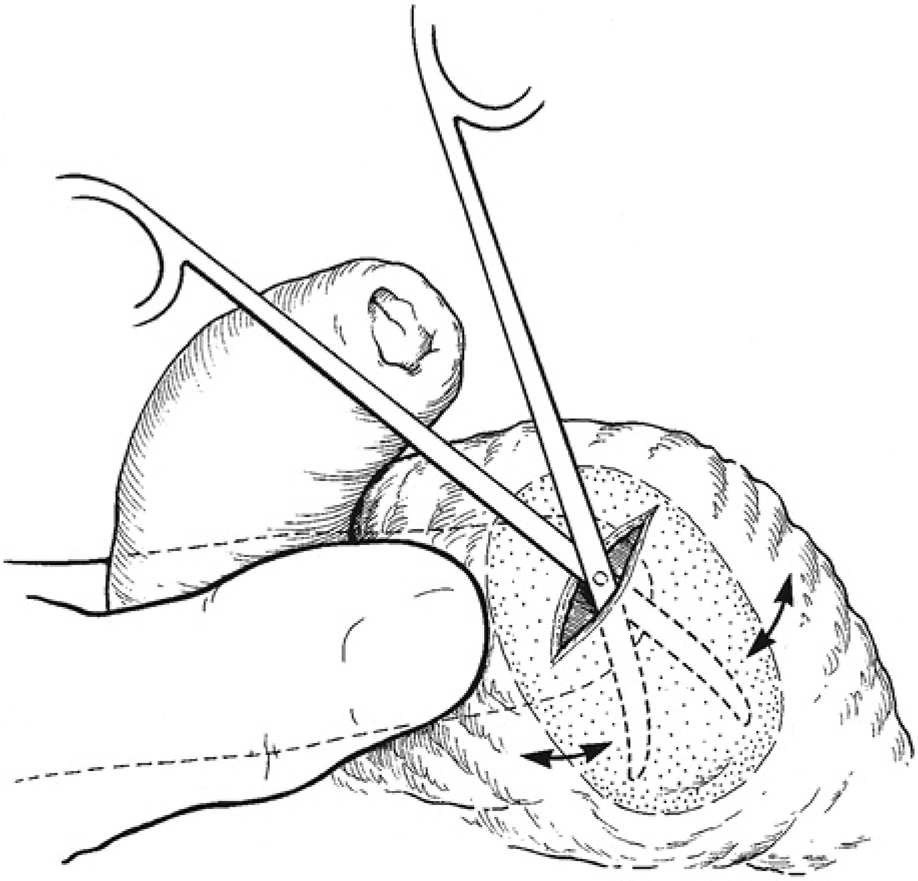
75.7

A finger is introduced through the incision and down to the scrotum, breaking down any fascial layers near the neck of the scrotum so that the tip of the index finger can reach the mid-scrotum. The scrotal skin is immobilized between the index finger internally and the thumb externally. A scrotal incision is then made, going just through the skin but not through the deeper tissues (**Figure 75.8**). This incision can be either horizontal (the author’s preference) or midline in the scrotal septum. Horizontal incisions are associated with less bleeding.



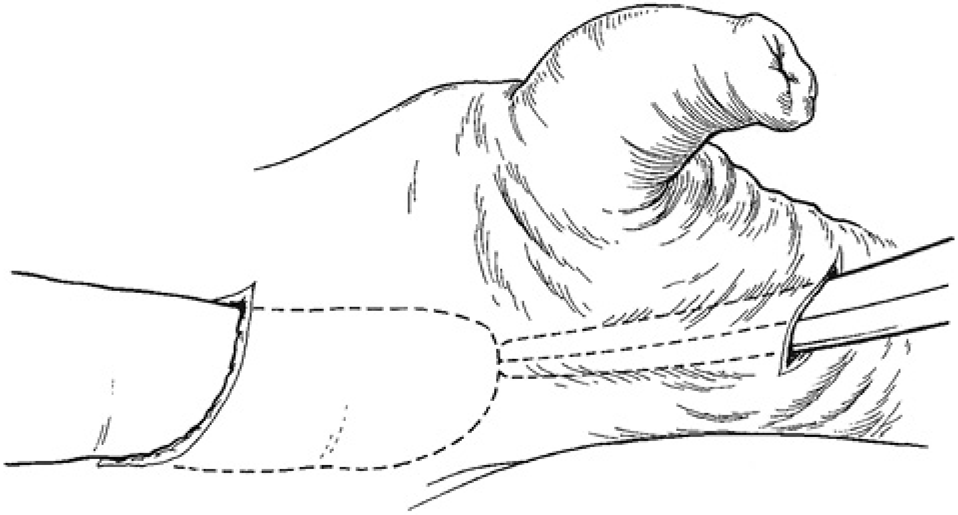
75.8

Fine artery forceps or scissors are used to develop a subcutaneous pouch just deep to the dartos muscle while the index finger is still inside the scrotum (**Figure 75.9**). This should be developed inferiorly more than superiorly, so that the external incision is placed near the cranial end of this subcutaneous pouch. Any bleeding is controlled by diathermy before proceeding further. This is an important step, as a scrotal hematoma will inevitably become infected and lead to wound breakdown.



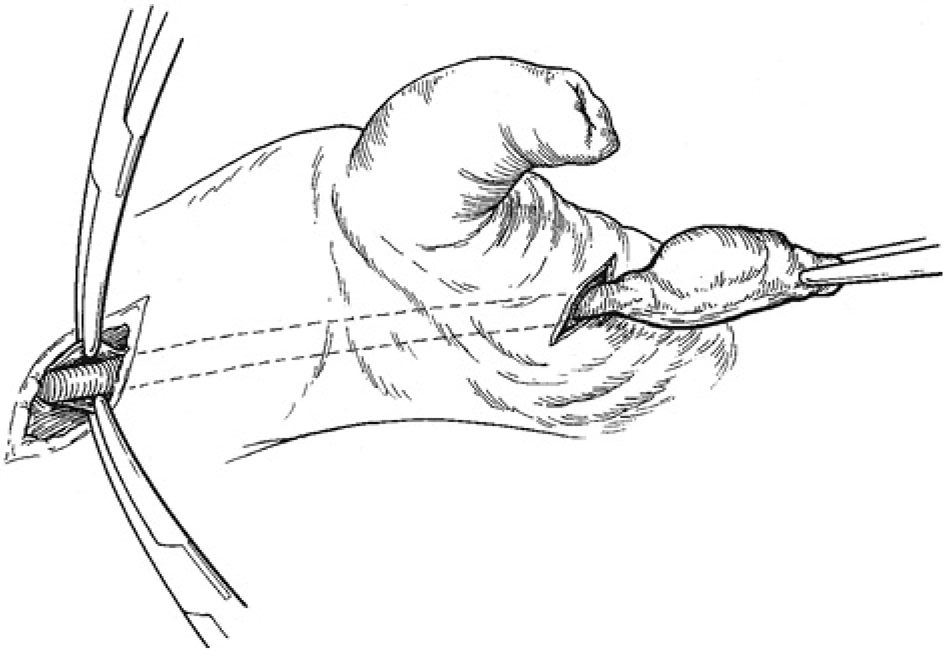
75.9

Artery forceps are placed through the incision in the scrotum and pressed against the index finger internally (**Figure 75.10**). The index finger then guides the artery forceps back to the inguinal incision where the tip of the artery forceps is pushed through any residual fascia.



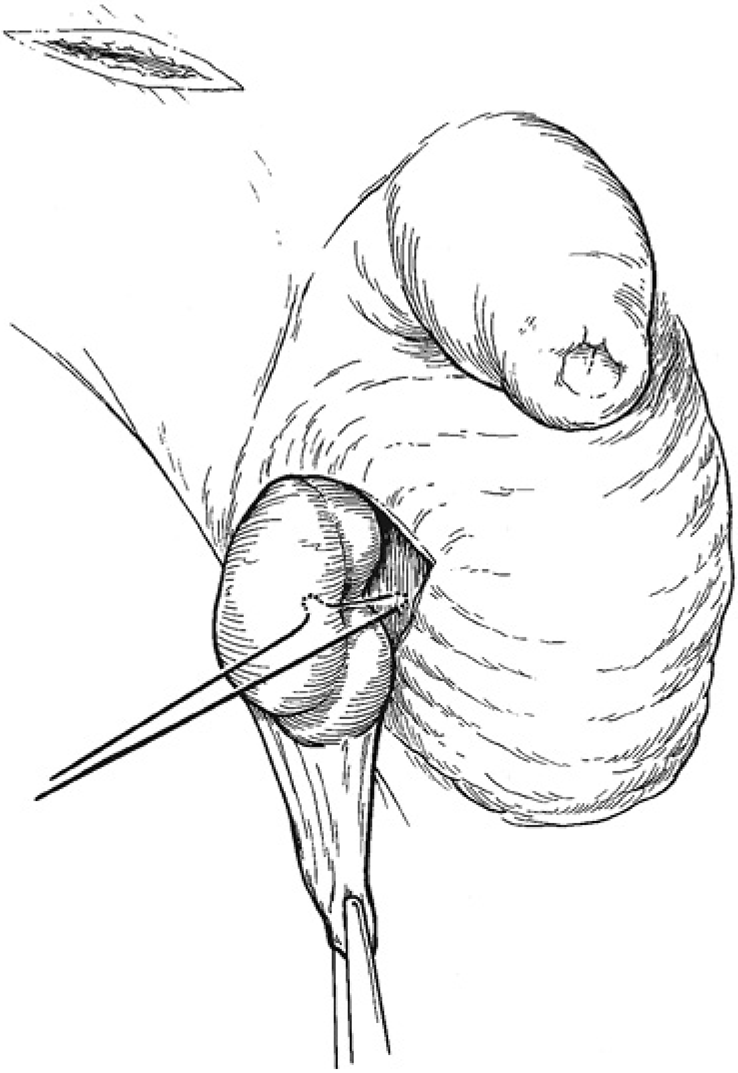
75.10

The artery forceps grasp the gubernacular attachments of the mobilized testis and, making sure that the cord structures are not twisted, the testis is then drawn gently down through the “buttonhole” of the subdartos fascia and delivered through the scrotal incision (**Figure 75.11**).



75.11

If the hole through the fascial planes between the two incisions is not too large, the testis will sit comfortably in the scrotal pouch like a button through a buttonhole (**Figure 75.12**). If there is any concern that the testis may retract through a larger defect, it can be anchored to the midline scrotal septum with a 3/0 absorbable suture through the tunica albuginea, but not through the body of the testis. Many surgeons deliberately open the tunica vaginalis so that the anatomy of the testis can be defined precisely and any testicular appendages can be excised at this time. The testis, epididymis, and adjacent coverings can then be placed in the subcutaneous scrotal pouch using blunt forceps. The scrotal incision is closed with a 4/0 subcuticular absorbable suture.



75.12

The operation is completed by reconstitution of the external oblique aponeurosis with continuous or interrupted 3/0 absorbable suture and closure of the inguinal wound.

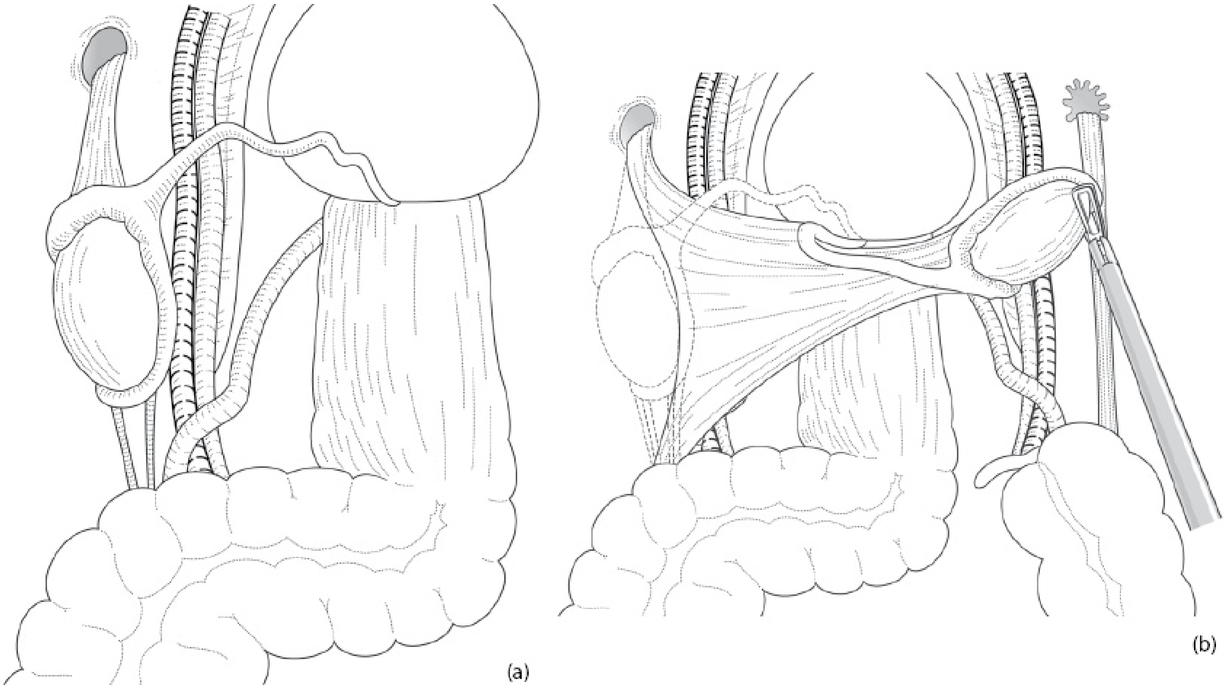
Scrotal orchidopexy

An upper scrotal incision can be made to deliver the testis where under anesthesia the testis can be manipulated into the scrotum and held there by the assistant. The rest of the operation is similar to standard orchidopexy, except that once the testis is delivered through the wound and put on traction with an artery forcep, the fat and then the external spermatic fascia needs to be stripped off the cord first. Mobilization up to the canal is often sufficient without opening it, especially for acquired cryptorchidism. If there is inadequate length, an inguinal incision with canalicular mobilization can be done as described above.

Laparoscopic orchiopexy

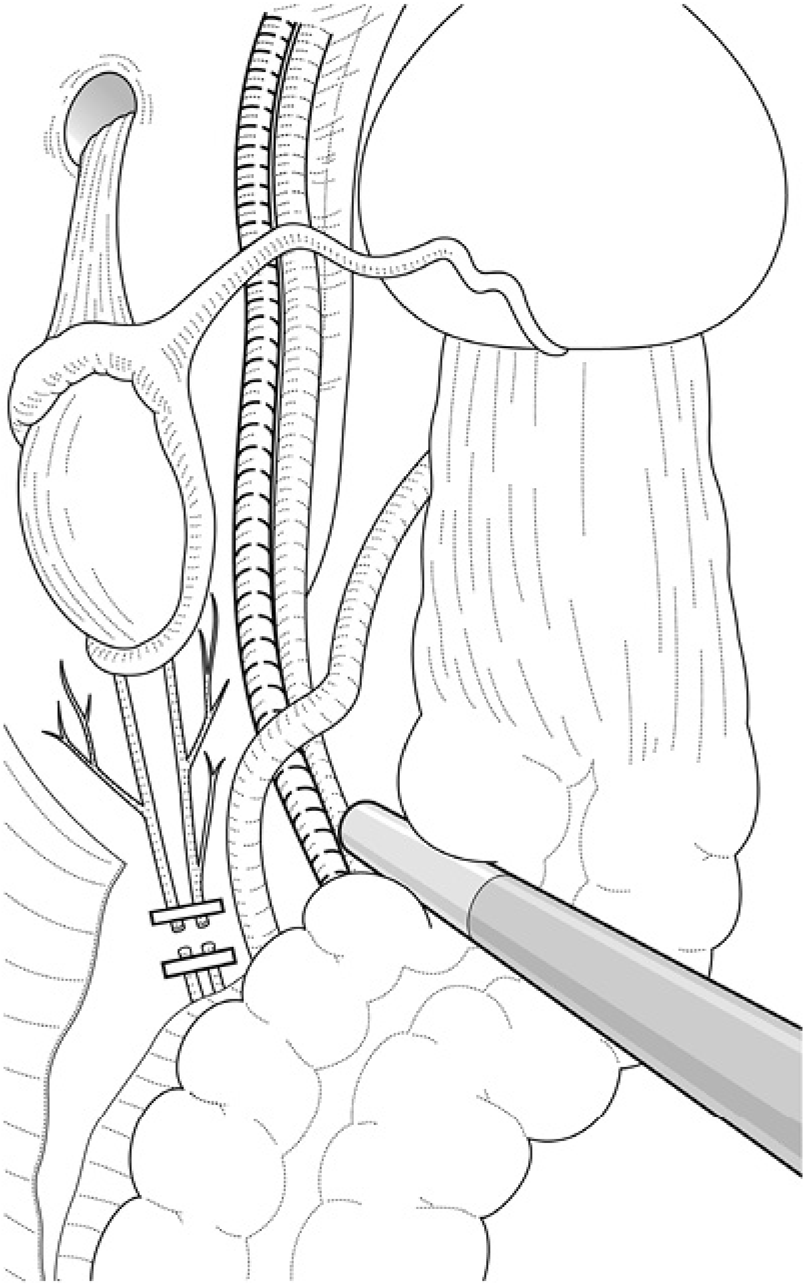
Diagnostic laparoscopy (as described in Chapter 27) is carried out for impalpable testes. This will identify the *vanishing testis*, where the internal inguinal ring is closed and the proximal testicular vessels appear from beneath the colon but are then blind-ending. The vas deferens may also be blind-ending, or exit through the closed inguinal ring. Some surgeons may then open the scrotum to remove the atrophic testicular nubbin (following presumed perinatal torsion), but this is controversial.

When the testis is intra-abdominal, it is usually close to the internal ring (**Figure 75.13a**). One-stage orchidopexy is straightforward if the testis can be dragged to the *opposite* internal ring with graspers, indicating adequate length of cord structures (**Figure 75.13b**).



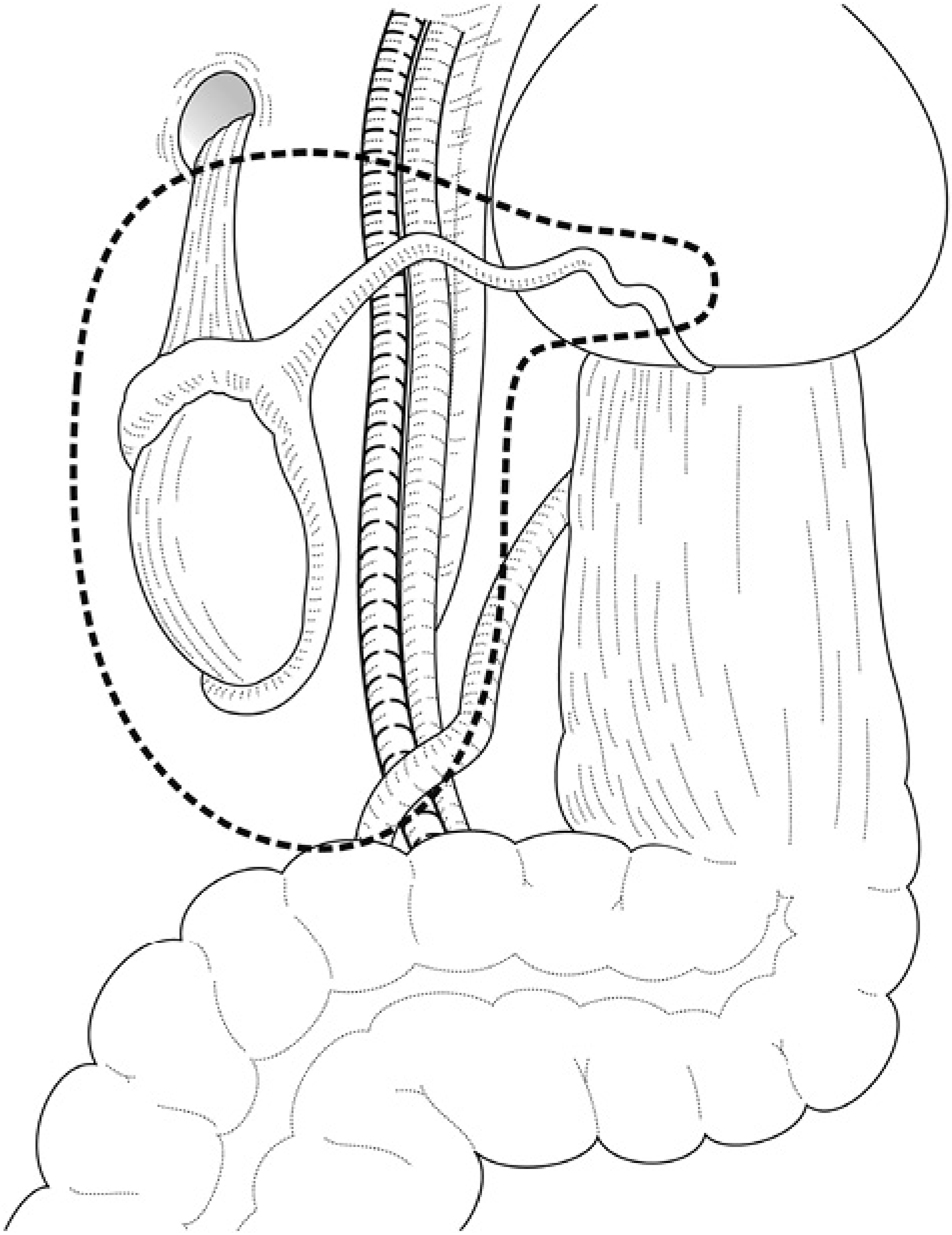
75.13a,b

For high intra-abdominal testes, the colon is mobilized to expose the proximal gonadal vessels far enough away from the testis to preserve any collateral vessels. The spermatic vessels are then clipped or ligated (**Figure 75.14**). This concept of ligation (and then division) of the testicular vessels was first described as part of the (open) Fowler–Stephens operation. The testis could then be mobilized satisfactorily only on the vas (and accompanying vasal vessels) to allow scrotal placement. Ransley modified it by separating the ligation phase and subsequent mobilization to reduce the risk of testicular ischemia.



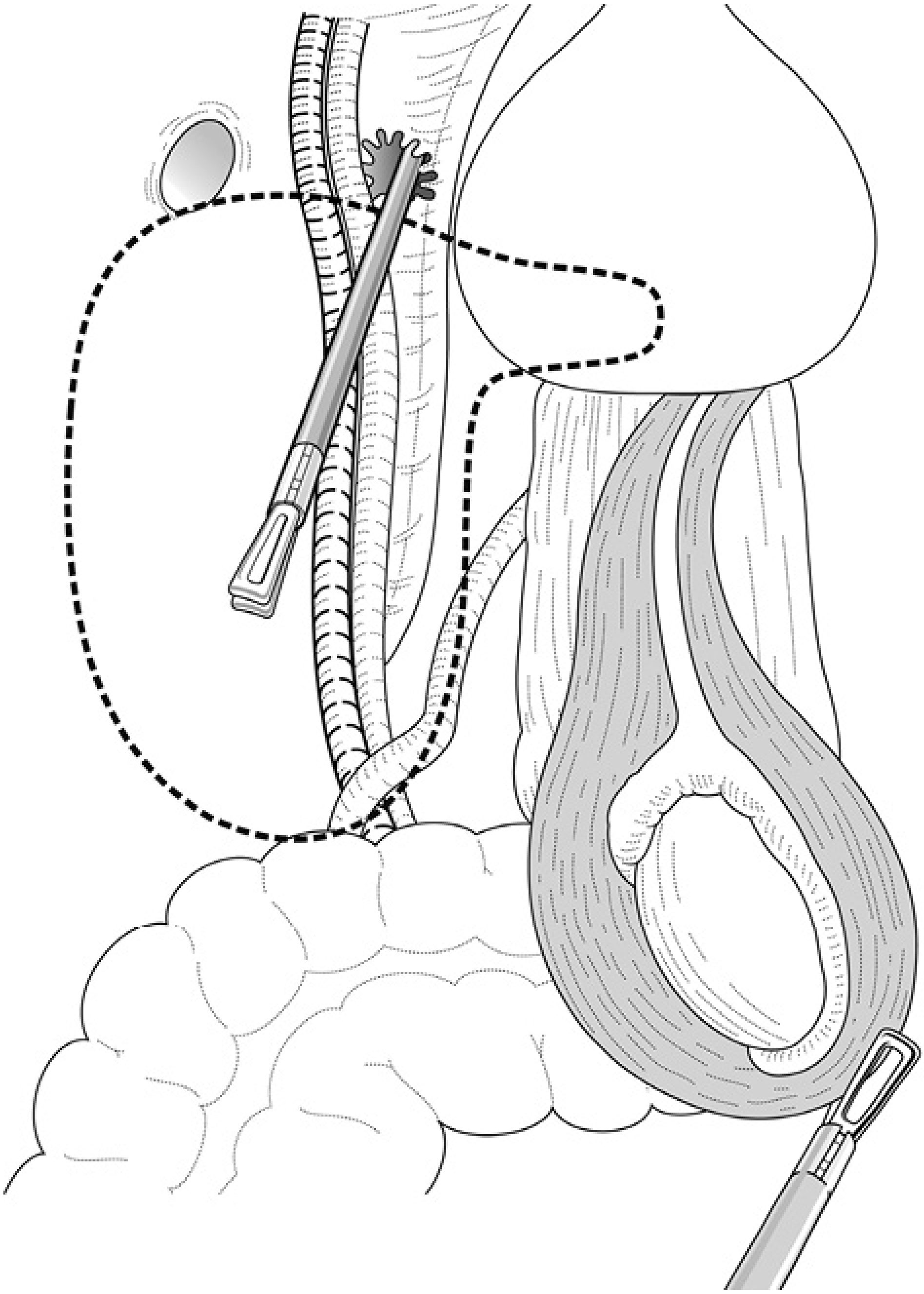
75.14

Six months later, after spermatic vessel clipping, or at the first laparoscopy if length is adequate, the peritoneum is divided in a tennis-racquet shape around the testis and vas deferens to mobilize the testis on its blood supply (**Figure 75.15**).



75.15

The scrotum is opened through a small incision and a grasper is pushed up into the abdomen through the external inguinal ring (and usually between the inferior epigastric vessels laterally and the obliterated umbilical artery medially) (**Figure 75.16**). It is important to ensure the tunnel created is stretched up enough to allow the testis to be pulled down. This is now grasped and pulled gently down to the scrotum, ensuring no twisting of the spermatic vessels. The operation is concluded by scrotal fixation as before.



75.16

COMPLICATIONS

Wound infection and hematoma are the two most common complications. Hematoma can be avoided by meticulous hemostasis with diathermy at the time of surgery. Wound sepsis can be avoided by the placement of waterproof dressings on both incisions, which remain in place for at least a week. The risk of testicular atrophy, which is determined at 6 months after surgery, should be less than 5%. There is a small risk of retraction of the testis into the groin, particularly if there is significant sepsis or hematoma, or postoperative trauma.

FURTHER READING

Fowler R, Stephens FD. The role of testicular vascular anatomy in the salvage of high undescended testes. *Aust N Z J Surg* 1959; 29: 92–106.

Hutson JM, Thorup J. Evaluation and management of the infant with cryptorchidism. *Curr Opin Pediatr* 2015; 27: 520–4.

Hvistendahl GM, Poulsen EU. Laparoscopy for the impalpable testes: Experience with 80 intraabdominal testes. *J Pediatr Urol* 2009; 5: 389.

Kravarusic D, Freud E. The impact of laparoscopy in the management of non-palpable testes. *Pediatr Endocrinol Rev* 2009; 7: 44.

Pastuszak AW, Lipshultz LI. AUA guideline on the diagnosis and treatment of cryptorchidism. *J Urol* 2014: 192: 346–9.

Steinbrecher H. The undescended testis: Working towards a unified pathway for 2014. *Arch Dis Child* 2014; 99: 397–8.

Toppari J, Rodprasert W, Virtanen HE. Cryptorchidism – disease or symptom? *Ann Endocrinol (Paris)* 2014; 75: 72–6.

Vikraman J, Hutson JM, Li R, Thorup J. The undescended testis: Clinical management and scientific advances. *Semin Pediatr Surg* 2016; 25: 241–8.

76

Testicular torsion

FEILIM L. MURPHY

HISTORY

While torsion of the testis itself must undoubtedly have been known about for millennia, attempts at surgical reduction were uncommon. One of the first concerns a young Parisian surgeon, Louis Delasiauve (1804–1893), who in 1839 explored the left groin of a 15-year-old boy with bilateral inguinal hernias. The groin had become swollen and painful and associated with absence of a scrotal testis. After an operation lasting 3 hours, he found a gangrenous undescended testis which had torted. This he removed.

INTRODUCTION

Torsion of the whole testis should be the first priority and consideration in any child with scrotal pain. There are two peaks of incidence during the perinatal period and between the ages of 11 and 25 years.

Neonatal torsion

Neonatal torsion accounts for up to 15% of cases of pediatric torsion. It is classically described as "extravaginal," implying that torsion occurs due to the failure of the tunica vaginalis to attach to the surrounding structures and is of the spermatic cord as a whole. However, evidence has been presented that some torsions in this period can be both intravaginal and extravaginal torsion.

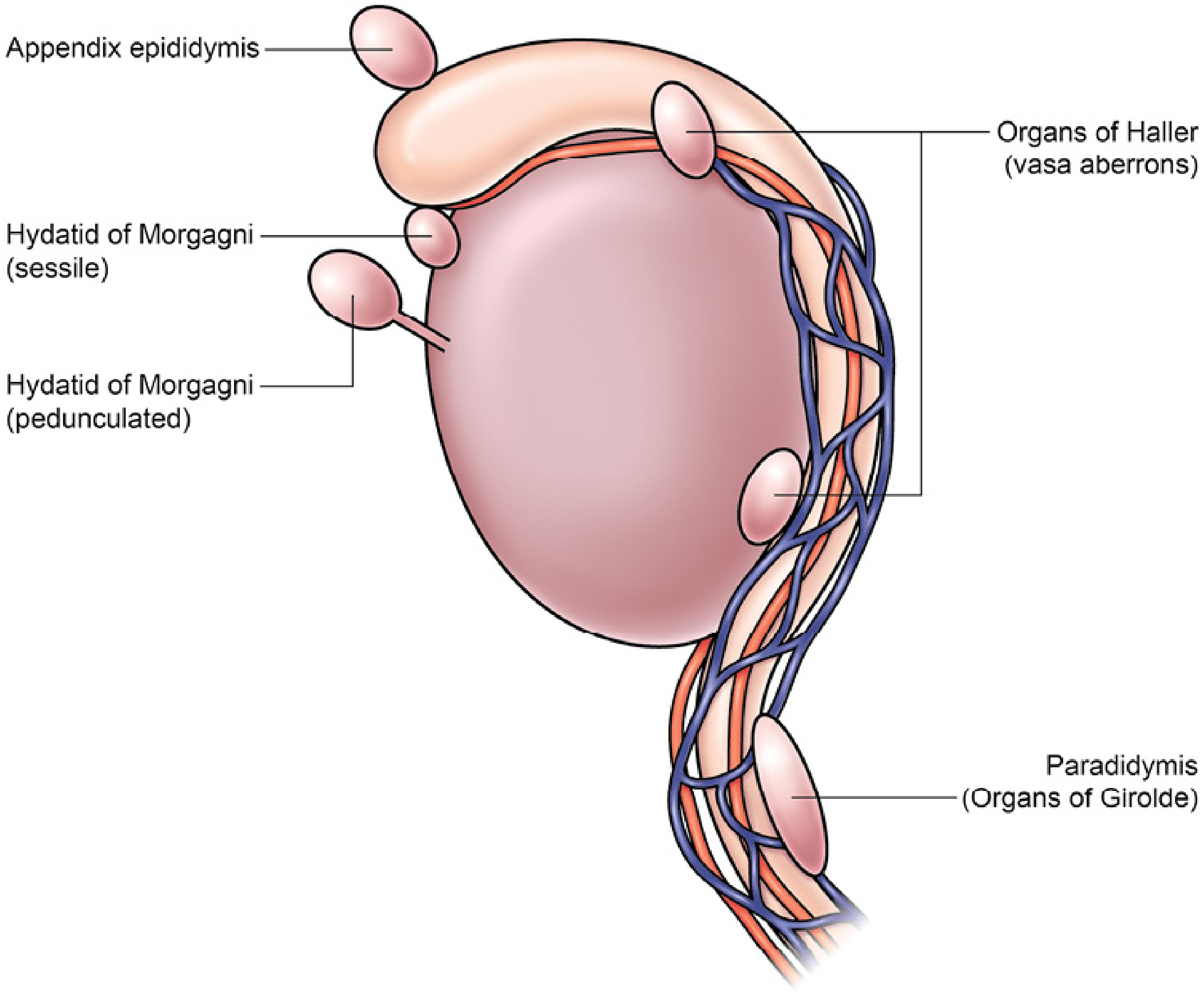
Due to reports of synchronous and asynchronous contralateral torsion and possible evidence of salvage of the postnatal testis, it is the author's view and recommendation that urgent exploration and contralateral fixation occurs.

Pathology of torsion

If the tunica vaginalis invests the whole of the epididymis and the distal part of the spermatic cord, the testis is in effect suspended in the scrotal cavity like a bell clapper and therefore is free to rotate within the tunica vaginalis in an intravaginal fashion.

DIFFERENTIAL DIAGNOSIS

Acute scrotal pain can also be due to torsion of one of the minor testicular appendages, and very rarely remnants of both Wolffian and Müllerian ancestry (**Figure 76.1**).


Appendix epididymis
Organs of Haller
(vasa aberrons)
Hydatid of Morgagni
(sessile)
Hydatid of Morgagni
(pedunculated)
Paradidymis
(Organs of Girolde)

76.1

Torsion of the testicular appendages

Torsion of the testicular appendages classically presents at 3–11 years. Careful palpation will reveal an exquisitely tender nodule which is usually associated with the upper pole of the testis while the body of the testis is not tender. On transillumination, a dark nodule or dot can be seen. If diagnosis of a torted appendix testis is made clinically, hospital observation or surgery are not mandatory.

Acute epididymo-orchitis

Unilateral epididymo-orchitis is rare in children but in these it may be associated with urinary tract infections and anomalies of the urinary tract.

Mumps

This rarely occurs before puberty, is usually bilateral, and tends to appear 3–7 days after the onset of parotitis. Elevated salivary amylase can be confirmatory for parotid involvement and real-time polymerase chain reaction is available in some centers.

Idiopathic scrotal edema

This is defined by erythema and swelling of the scrotal skin into the groin, perineum, and/or the base of the penis. Patients are usually aged 3–6 years and do not usually complain of pain. However, the skin may be tender but testis and cord are normal. The diagnosis can be made clinically and reassurance is all that is required.

OPERATION

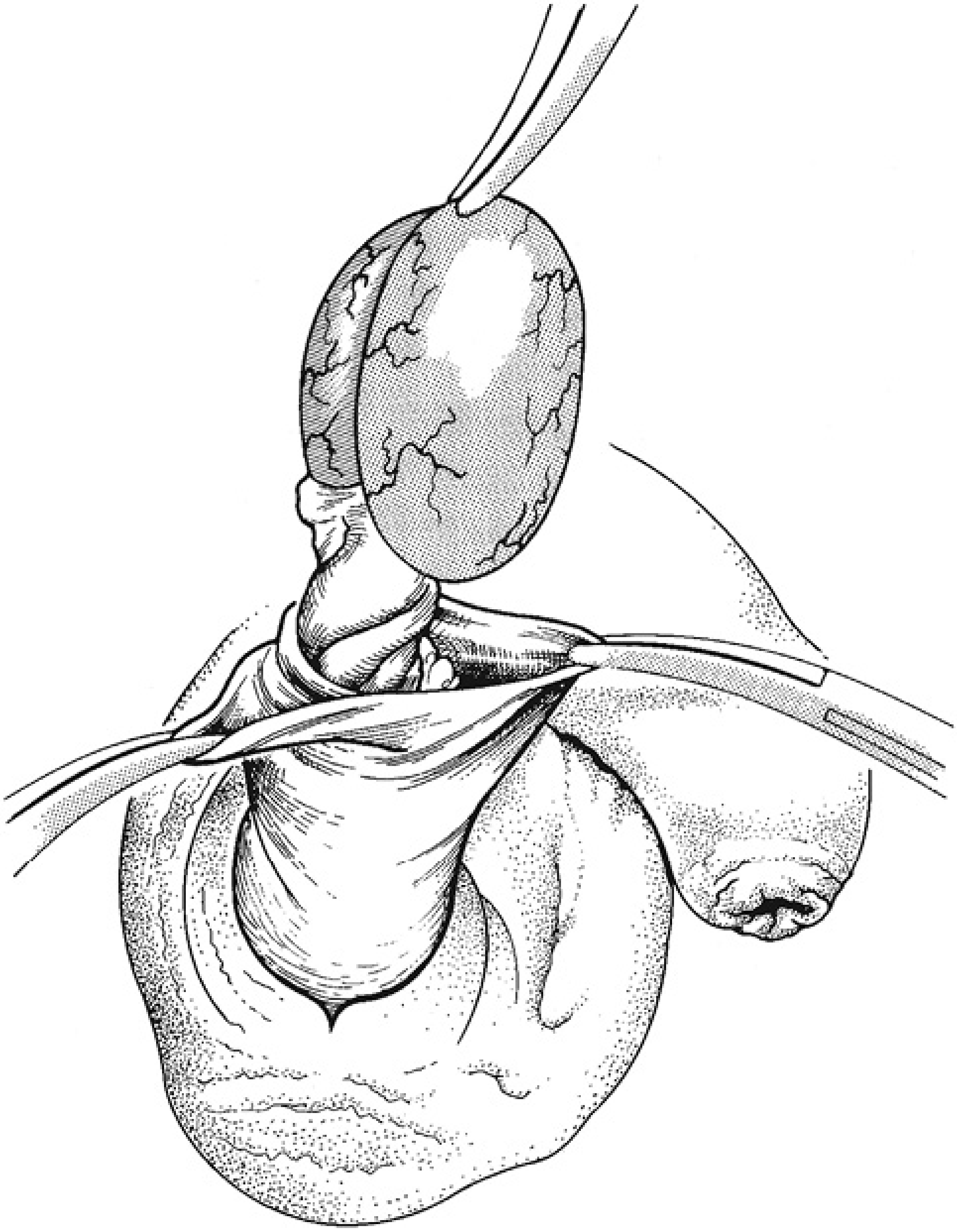
Timing

In most cases the diagnosis of the acute testicular torsion can be made clinically. Once the diagnosis is suspected, urgent surgery is required and any delay will prejudice the survival of the testis.

Incision

The testis can be delivered from the scrotum through an incision over the longitudinal axis or obliquely in the line of the rugae or through the midline raphe. The latter allows exploration of both sides of the scrotum through the same incision.

The testis is delivered from the tunica vaginalis and the cord is untwisted in the appropriate direction (**Figure 76.2**). The tunica albuginea can be incised to assess viability and it can relieve the pressure within the tubules.



76.2

The untwisted testis should be wrapped in moist warm swabs and its color observed intermittently. While waiting to confirm whether perfusion can be re-established, the contralateral testis can be explored and fixed.

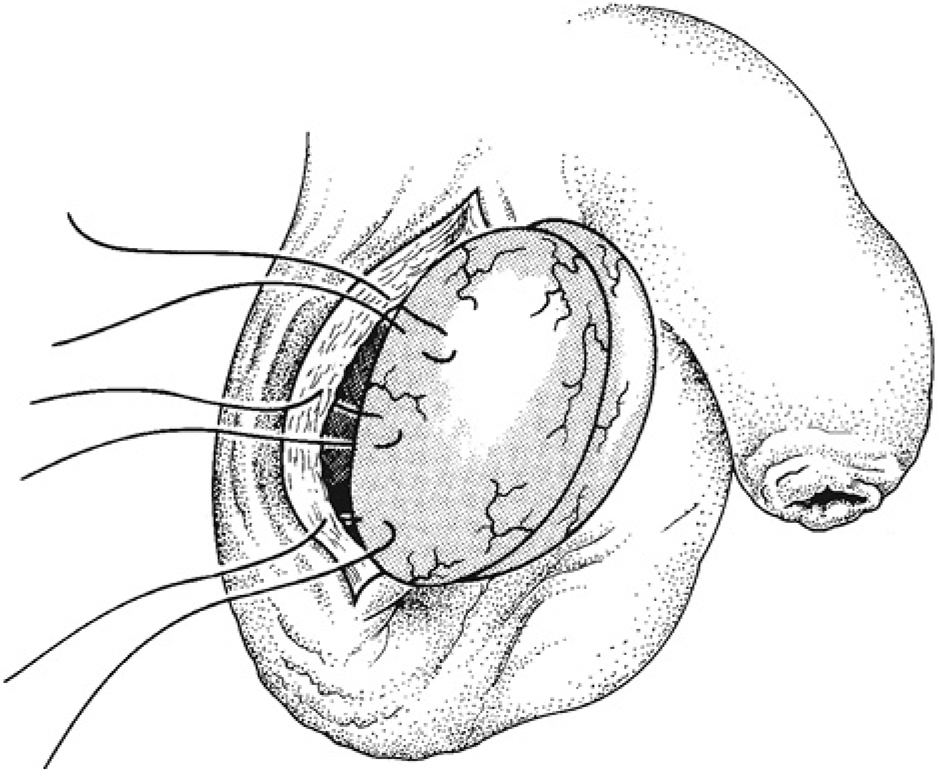
Conservation or removal

If the testis is completely black, necrotic, soft, and non-viable, it should be removed. The spermatic cord is ligated within the scrotum with an absorbable suture and the testis excised. If there is any question that some perfusion might be re-established, the testis should be conserved. The contralateral testis must always be fixed.

Fixation of the testis

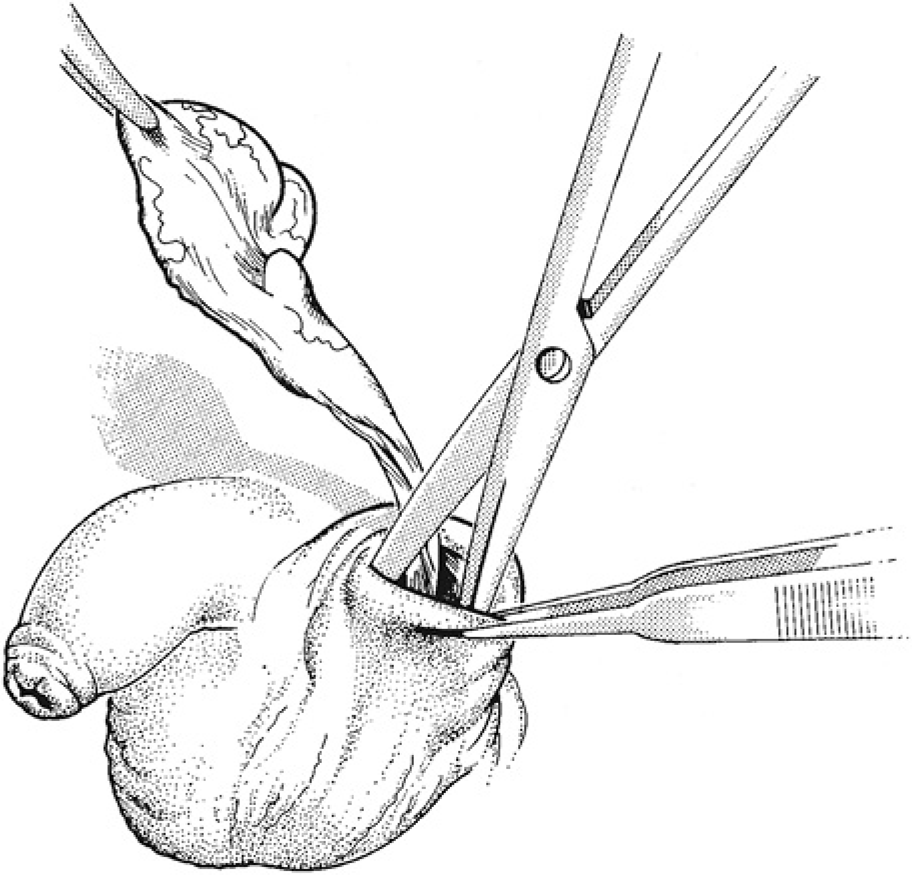
Two methods of fixation are commonly used:

The testis can be everted from the tunica vaginalis and fixed with three 6/0 monofilament sutures placed within the tunica albuginea and the lateral wall of the scrotum at the upper and lower poles and the equator (**Figure 76.3**). There is concern that the use of the absorbable sutures may allow torsion to occur at a future while non-absorbable can be associated with chronic pain.



76.3

The testis is retracted superiorly. Fine-toothed forceps lift the edge of the wound and with blunt dissection an adequate “sub-dartos” scrotal pouch is formed (**Figure 76.4**). The testis, which is still everted from the tunica vaginalis, is placed in the scrotal pouch.



76.4

The wound is closed continuously or interrupted with 4/0 to 6/0 absorbable sutures. If an incision is made in the midline, a layered closure is needed to recreate the septum.

POSTOPERATIVE CARE

Patients with true torsion should be followed up to monitor testicular size and ensure growth.

FURTHER READING

Frank JD, O'Brien M. Fixation of the testis. *Br J Urol* 2002; 89: 331–3.

Kyriaziz ID, Dimopoulos J, Sakellaris G, et al. Extravaginal testicular torsion: A clinical entity with unspecified surgical anatomy. *Int Braz J Urol* 2008; 34: 617–23.

Soccorso G, Ninan GK, Rajimwale A, Nour S. Acute scrotum: Is the scrotal exploration the best management? *Eur J Pediatr Surg* 2010; 20: 313–15.

77

Circumcision, meatotomy, meatoplasty, and preputioplasty

RACHEL HARWOOD AND SIMON E. KENNY

PRINCIPLES AND JUSTIFICATION

Indications

Most circumcisions are performed for non-medical reasons, namely religious beliefs or cultural practice. Jewish boys are circumcised on the eighth day of life, and Muslim boys are usually circumcised during childhood. Most of these religious circumcisions are performed in community settings. Additionally, many boys are circumcised in the neonatal period, particularly in the United States for cultural reasons.

The medical indications for circumcision include pathological phimosis, usually related to balanitis xerotica obliterans (BXO) and prophylactic circumcision in boys at risk of recurrent urinary tract infections due to an abnormal urinary tract. A relative indication is recurrent severe balanoposthitis. It is the authors' experience that this is mostly a self-limiting phenomenon, resolving with non-operative interventions (e.g. topical steroids).

Physiological phimosis is a self-resolving condition that usually requires reassurance. Ballooning of the prepuce during micturition is of no consequence and not an indication for circumcision. If physiological phimosis is symptomatic, topical steroid therapy can be effective and should be tried prior to considering surgery.

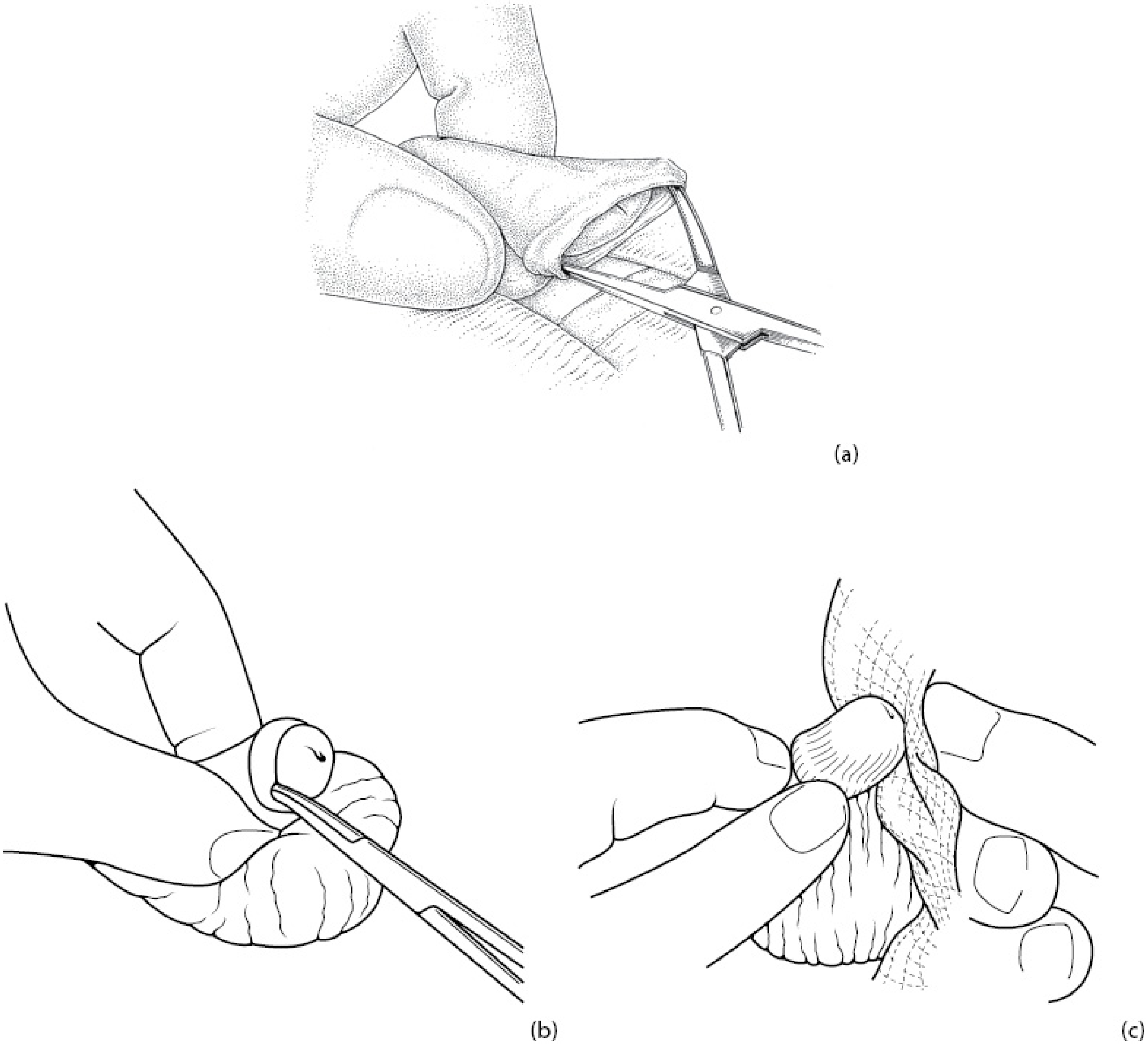
Contraindications for circumcision include hypospadias and buried penis. Meatal stenosis can arise following circumcision, particularly in boys with BXO and those who have a circumcision while still in diapers. Most of these patients can be treated by gentle meatal dilatation or meatotomy.

An alternative to circumcision is preputioplasty and intradermal steroid injection. This permits preservation of the foreskin, although there is a risk of recurrent phimosis in boys with BXO. Many boys opt for preputioplasty and the procedure should be discussed with boys and their parents as part of the preoperative decision-making process.

OPERATIONS

Sleeve circumcision

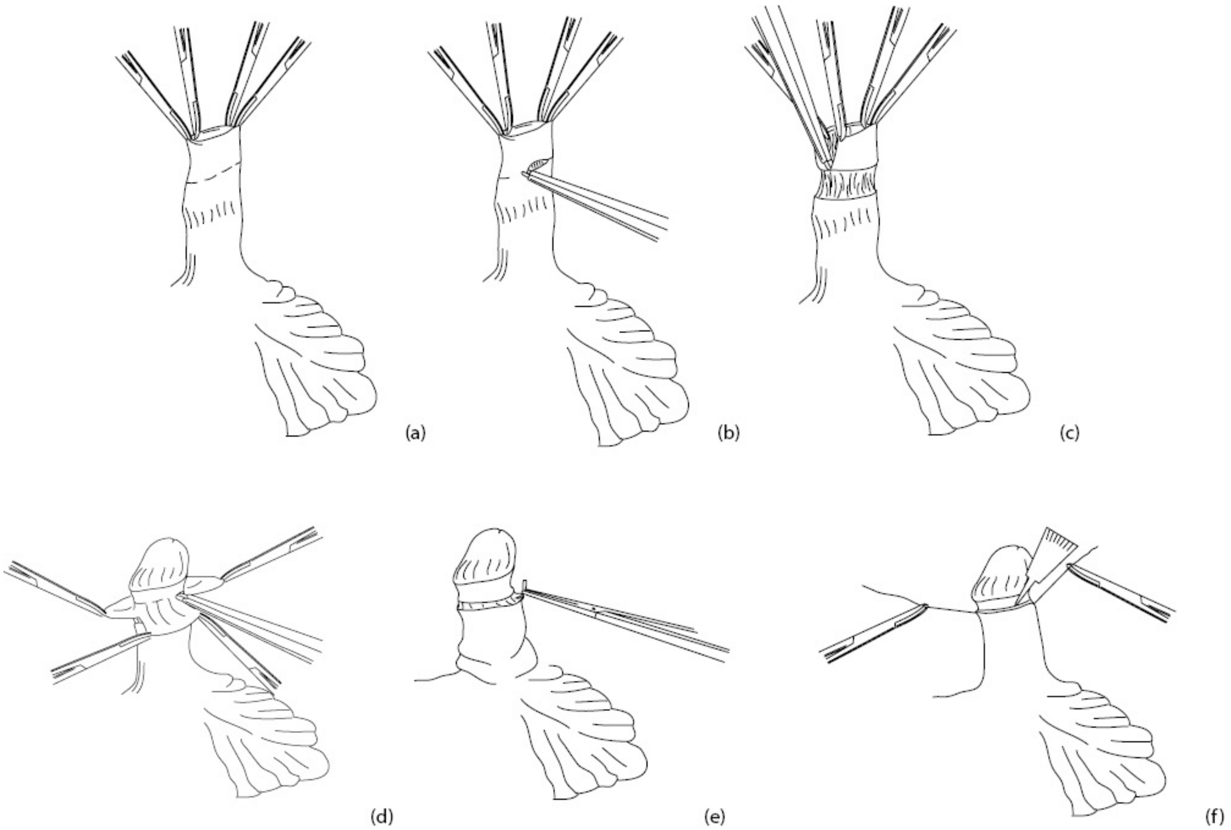
There are many ways in which to perform this procedure. The following is the author's preferred method. The phimosis is carefully dilated using artery forceps (avoiding the glans) to permit retraction of the foreskin and division of preputial adhesions (**Figure 77.1a–c**).



77.1a–c

In severe BXO or phimosis, a dorsal slit may be required in order to retract the prepuce after applying artery forceps as described below. A marker pen is used to mark the position of the external preputial incision by observing the coronal ridge of the glans under the foreskin. Care needs to be taken in ensuring that neither excessive nor insufficient penile shaft skin is removed. Similarly, the inner preputial incision is marked with a marker pen leaving an adequate cuff of inner prepuce below the glans penis. This can be performed after the external skin has been incised.

Four artery forceps are applied to the prepuce – two ventrally and two dorsally – and upward traction applied (**Figure 77.2a**). The external penile shaft skin is incised along the marker line using bipolar diathermy forceps or scissors (**Figure 77.2b**). A scalpel can be used but this increases intra- and postoperative blood loss. The prepuce is divided ventrally and dorsally between the artery forceps to the level of the outer incision (**Figure 77.2c**). This maneuver effectively splits the prepuce into two halves, which aids accurate incision of the inner prepuce along the previously marked line using bipolar forceps (**Figure 77.2d**).



77.2a–f

The outer skin is retracted, and any bleeding points are fastidiously coagulated with bipolar diathermy. Ligatures may be used if bipolar diathermy is unavailable. The wound is closed with an interrupted absorbable suture, such as 5/0 Vicryl□ rapide or Monocryl□. Sutures should be placed close (<1 mm) to the wound edges to avoid unsightly scarring (**Figure 77.2e**). Tissue glue is an effective and quick alternative to suturing favored by the authors after placement of a stay suture ventrally and dorsally to aid approximation of the wound edges (**Figure 77.2f**).

POSTOPERATIVE CARE

The application of chloramphenicol ointment twice daily for 7 days may help to prevent infection and stops underwear adhering to the penis. Dressings should be avoided as they stick to the wound and are painful to remove.

COMPLICATIONS

Circumcision is often regarded as a minor surgical procedure. However, there are probably more complications associated with this operation than with more complex urological procedures.

Hemorrhage

Parents and the child should be warned that some oozing is not uncommon but, if this is severe or persistent, they should return to hospital immediately. After circumcision, there is a 1% incidence of bleeding sufficient to warrant a return to theatre. Bleeding is more common following circumcision for BXO.

Infection

Infections after circumcision are not uncommon, with an incidence of around 5%. They generally respond well to oral antibiotics and regular bathing.

Urethrocutaneous fistulas

Fistula formation is a rare complication of circumcision and is usually seen at the level of the coronal sulcus. The most common causes are overzealous use of diathermy and a suture placed too deeply to stop bleeding from the frenulum. Surgical repair should be performed 6 months after the original circumcision.

Removal of too much or too little skin

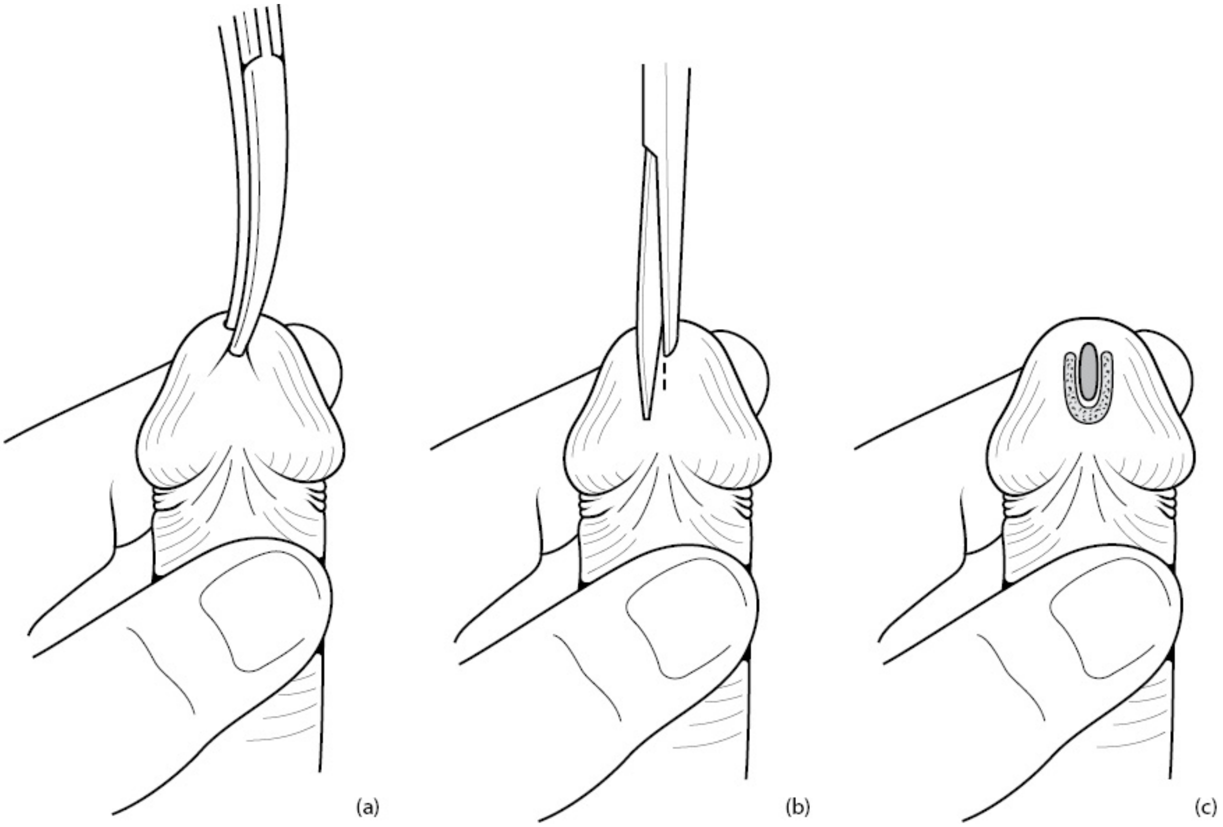
Removal of too little skin can cause annular scarring at the line of excision and recurrence of the phimosis. In such cases, a further circumcision is required. Removal of too much skin may give the appearance of a buried penis, but only rarely is skin grafting required to achieve a satisfactory appearance.

Meatal stenosis

Meatal stenosis is rare (<1%) following circumcision and the postoperative application of petroleum jelly for a few weeks appears to reduce the risk. It is more common in boys with BXO with glanular involvement and 1 : 5 boys will require a further meatal procedure following circumcision for BXO.

Meatotomy

In meatotomy (**Figure 77.3a–c**) the meatus is calibrated with a urethral sound to determine the severity of the stenosis. An artery forceps is used to crush the tissue ventrally through the meatus for 1 minute. The artery forceps are removed and the crushed tissue divided with iris scissors. Bleeding is rare, but it can be readily controlled with bipolar diathermy.



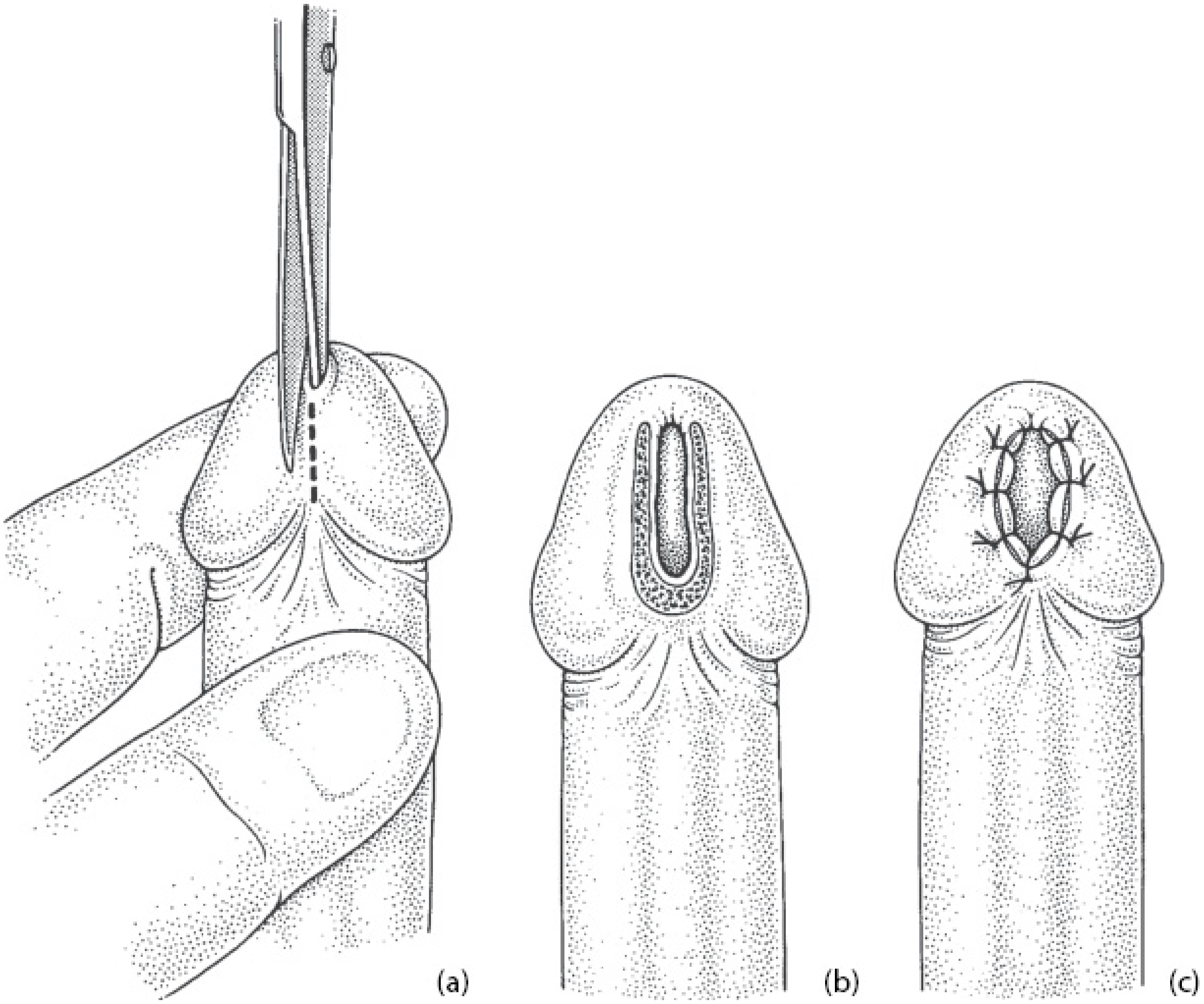
77.3a–c

COMPLICATIONS

The main complication of this procedure is recurrence of the meatal stenosis, which may require a meatoplasty.

Meatoplasty

A pair of iris scissors is used to make a generous ventral incision through the meatus (**Figure 77.4a and b**). The edges of the urethral mucosa are approximated to the adjacent skin with 7/0 or 6/0 absorbable sutures (**Figure 77.4c**).



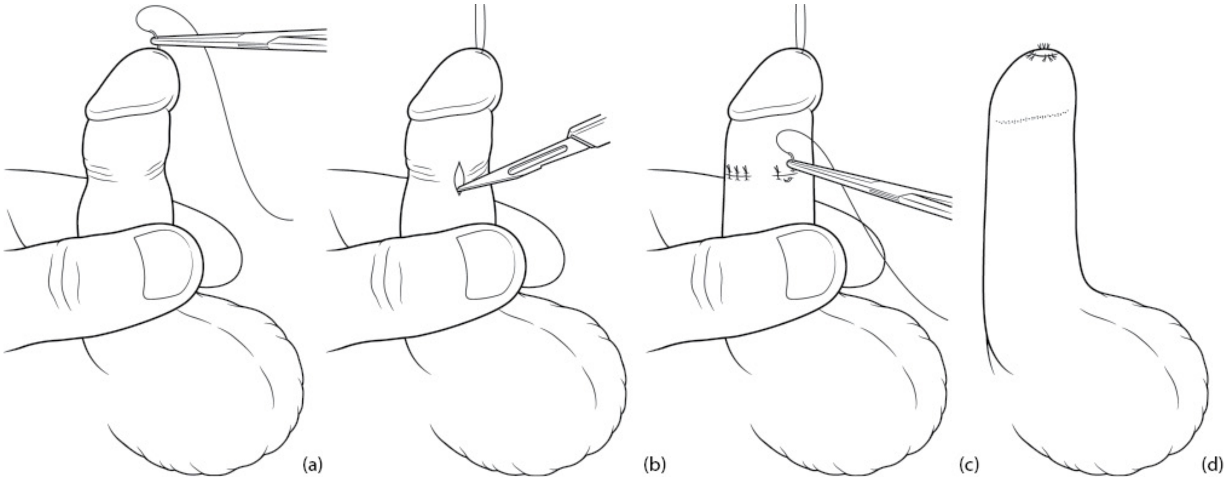
77.4a–c

COMPLICATIONS

This procedure creates a large meatus with the appearance of a glanular hypospadias. This may cause some deviation of the urinary stream, but it is unlikely to cause functional difficulties.

Preputioplasty

If possible, the phimotic foreskin is dilated, retracted and the glans cleaned, as with circumcision. The band of phimotic skin is divided longitudinally in three places: Dorsally and ventrolaterally on each side, through to the dartos fascia layer (**Figure 77.5a and b**). The incisions naturally lie transversely and increase the circumference of the prepuce. They are extended until the foreskin can be freely retracted and replaced. It is sometimes not possible to retract the prepuce over the glans without splitting it. In this situation, the prepuce should be retracted as far as possible and incisions made in the same positions until the prepuce can be retracted. The glans should be protected throughout.



77.5a–d

If BXO is present, triamcinolone (10 mg/mL) can be infiltrated subcutaneously around the prepuce. Bipolar diathermy should be used to ensure hemostasis. 5/0 interrupted Vicryl□ rapide or Monocryl□ is used to approximate the skin edges of each incision transversely prior to replacing the prepuce over the glans (**Figure 77.5c**). Chloramphenicol can be applied as with circumcision.

COMPLICATIONS

The main complication with preputioplasty is the recurrence of phimosis which may require circumcision in the future. With BXO, this risk is about 20%. The risk can be reduced by asking that boys retract and replace their foreskin twice a day, starting 2 weeks after preputioplasty. If phimosis is recurring, a topical steroid cream can be beneficial.

Boys and their families should be carefully counseled about the choice between circumcision and preputioplasty prior to undertaking either procedure.

FURTHER READING

Green PA, Bethell GS, Wilkinson DJ, et al. Surgical management of genitourinary lichen sclerosus et atrophicus in boys in England: A 10-year review of practices and outcomes. *J Pediatr Urol* 2019; 15: 45.

Liu J, Yang J, Chen Y, et al. Is steroid therapy effective in treating phimosis? A meta-analysis. *Int Urol Nephrol* 2016; 48: 335–42.

Singh-Grewal D, Macdessi J, Craig J. Circumcision for the prevention of urinary tract infection in boys: A systematic review of randomised trials and observational studies. *Arch Dis Child* 2005; 90: 853–8.

Wilkinson DJ, Lansdale N, Everitt LH, et al. Foreskin preputioplasty and intralesional triamcinolone: A valid alternative to circumcision for balanitis xerotica obliterans. *J Pediatr Surg* 2012; 47: 756–9.

78

Varicocele

CHARLOTTE V. MELLING, DAVID J.B. KEENE, AND RAIMONDO M. CERVELLIONE

HISTORY

Varicocele is likely a consequence of bipedal walking and upright motion, which changed forever the mechanics of deoxygenated blood flow from the scrotum to the heart. Coupled with rapid peripubertal linear growth in adolescence, anti-gravitational forces can overcome venous valvular competence, resulting in a varicocele. Surprisingly astute reports from 1st century CE Greek scholar, Celsus, describe how "the veins are swollen and twisted over the testicle, which becomes smaller."

PRINCIPLES AND JUSTIFICATION

Varicocele is the abnormal dilatation and tortuosity of the internal spermatic veins within the pampiniform plexus. Clinically significant varicocele results from continuous or spontaneous rather than Valsalva-induced spermatic venous reflux. The condition is common is adolescents (10–20%) and has a similar prevalence to that of adult males (15%). They can cause pain or discomfort and limit sporting activities. Importantly, varicocele contributes to sub-fertility, and is more common in men seeking infertility treatment (25–35%). The impact on paternity is less clear, as up to 85% of men with varicocele still father children.

Anatomy and pathophysiology

Collecting venules from the testes and epididymis unite posteriorly to form the pampiniform plexus, which ascends within the inguinal canal before emerging through the deep ring as three or four larger veins eventually to form a single vein. As they ascend the posterior abdominal wall, they coalesce further, entering the inferior vena cava on the right and the left renal vein on the left. The venous anatomy between the internal inguinal ring and the left kidney can be very variable, and complex venous malformations and duplications have been demonstrated on the left side (see Nagappan et al., 2015, in "Further reading"). Varicocele is almost universally left-sided, due to several factors:

- The 90-degree angle of the left testicular vein junction with the left renal vein results in valvular incompetence
- Compression of the left renal vein between the aorta and the superior mesenteric artery ("nutcracker phenomenon")
- The presence of venous malformations

Grading

The Dubin and Amelar (1970) clinical grading of varicocele is the most widely used:

- *Grade 1:* Palpable only on Valsalva maneuver
- *Grade 2:* Palpable but not visible
- *Grade 3:* Palpable and visible

However, the most clinically important feature is the presence of continuous venous reflux, which can only be assessed with Doppler ultrasonography. For clinical purposes, the authors recommend to differentiate simply between Valsalva-induced reflux and spontaneous venous reflux. The latter is always associated with Grade 2 and Grade 3 varicocele.

INVESTIGATIONS

- The patient is examined in standing position and the varicocele grading is recorded.
- A scrotal Doppler ultrasound (US) is performed with the patient standing and venous reflux is recorded.
- Testicular volumes are recorded, preferentially with US. A 20% volume discrepancy between the two testes is considered significant as a sign of testicular growth arrest.
- Abdominal US is requested only in the presence of a right-sided varicocele (extremely rare) or the absence of venous reflux in a Grade 2–3 varicocele (suggesting an obstructive nature of the varicocele).
- Sperm analysis can be requested in post-pubertal boys as sperm concentration and forward motility are age-independent.

Indications for intervention

The asymptomatic varicocele without testicular growth arrest or impaired sperm parameters can be managed expectantly with 6–12 monthly reviews. Current European Association of Urology (EAU) guidelines recommend intervention in the presence of:

- Testicular growth arrest
- Additional testicular pathology
- Bilateral palpable varicocele
- Abnormal sperm parameters
- Symptomatic varicocele

Emerging evidence on the importance of impaired sperm parameters in adolescents, which may precede testicular growth arrest, should be carefully considered. The authors have previously shown that correction of varicocele in boys with asymmetrical testes can restore sperm parameters to normal in about 40%, and improve the remainder.

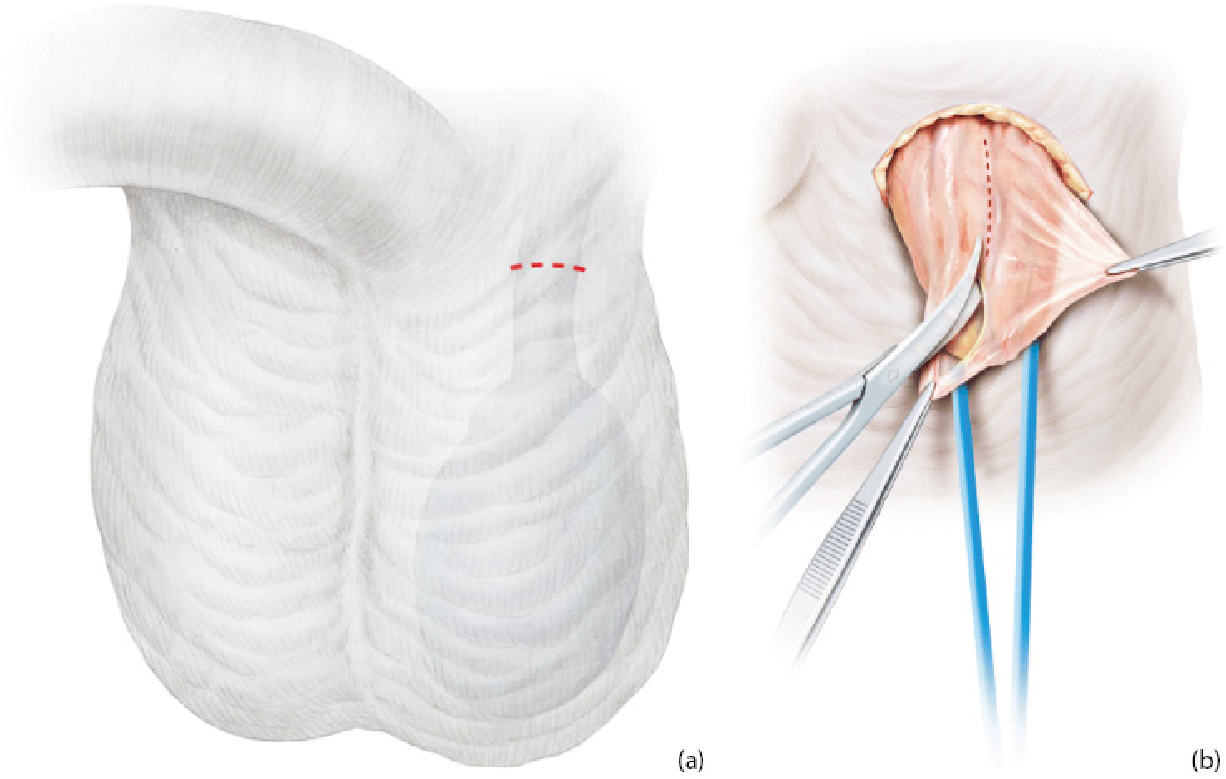
TECHNIQUES IN VARICOCELE MANAGEMENT

The management of varicocele remains highly variable in terms of surgical approach and methodology, but techniques include:

- Open inguinal or sub-inguinal surgery.
- Retroperitoneal laparoscopic selective ligation of the testicular vein(s) (Ivanissevich operation).
- Retroperitoneal mass ligation of vein(s), artery and lymphatics (Palomo operation).
- *Antegrade sclerotherapy*: A recent poll of UK pediatric urologists at the British Association of Paediatric Urologists (BAPU) 2018 Consensus Session indicated that 54% would recommend antegrade sclerotherapy for managing pediatric and adolescent varicocele.

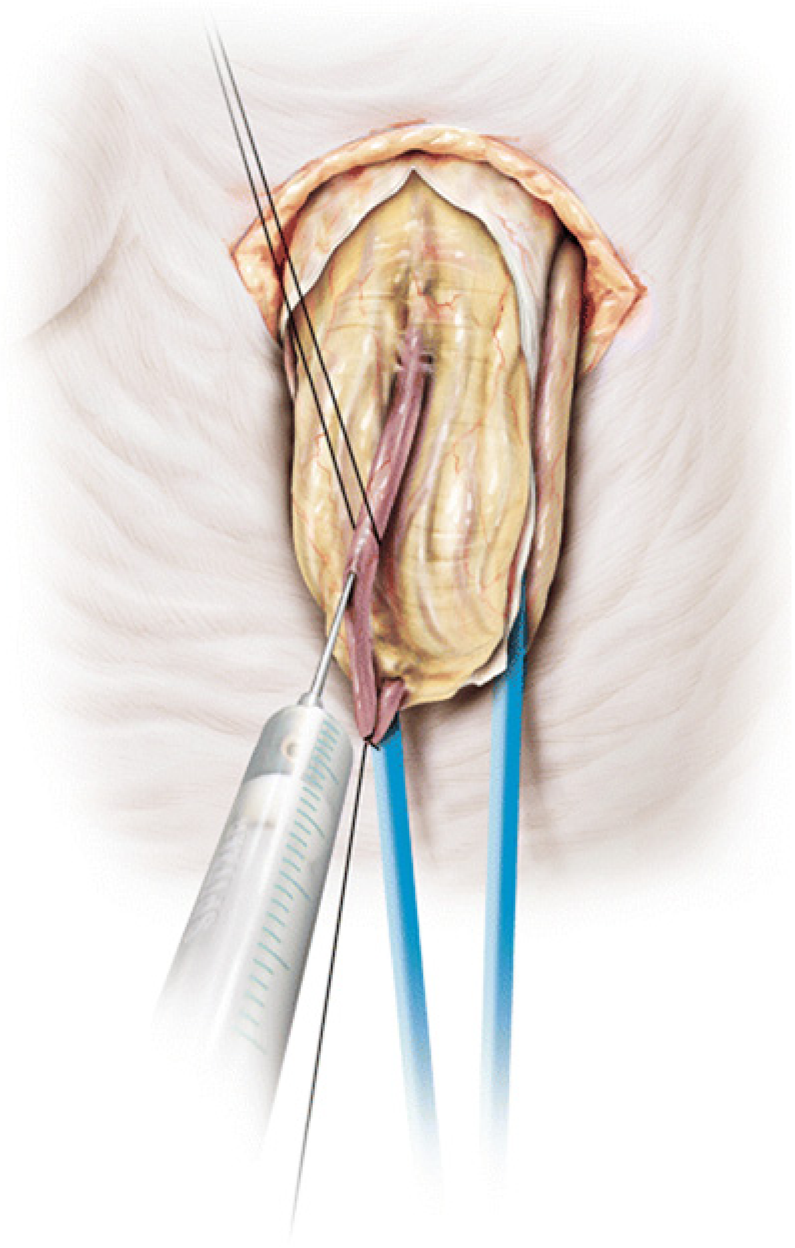
Antegrade sclerotherapy

This is performed under general anesthetic and the patient is positioned supine on a spinal extension table to enable X-ray screening up to the kidney. An upper scrotal incision is made over the spermatic cord (**Figure 78.1a**) and the external spermatic fascia is opened (**Figure 78.1b**). The pampiniform plexus is identified within the fat of the cord, and a single vein is isolated and slooped with 3/0 ties proximally and distally.



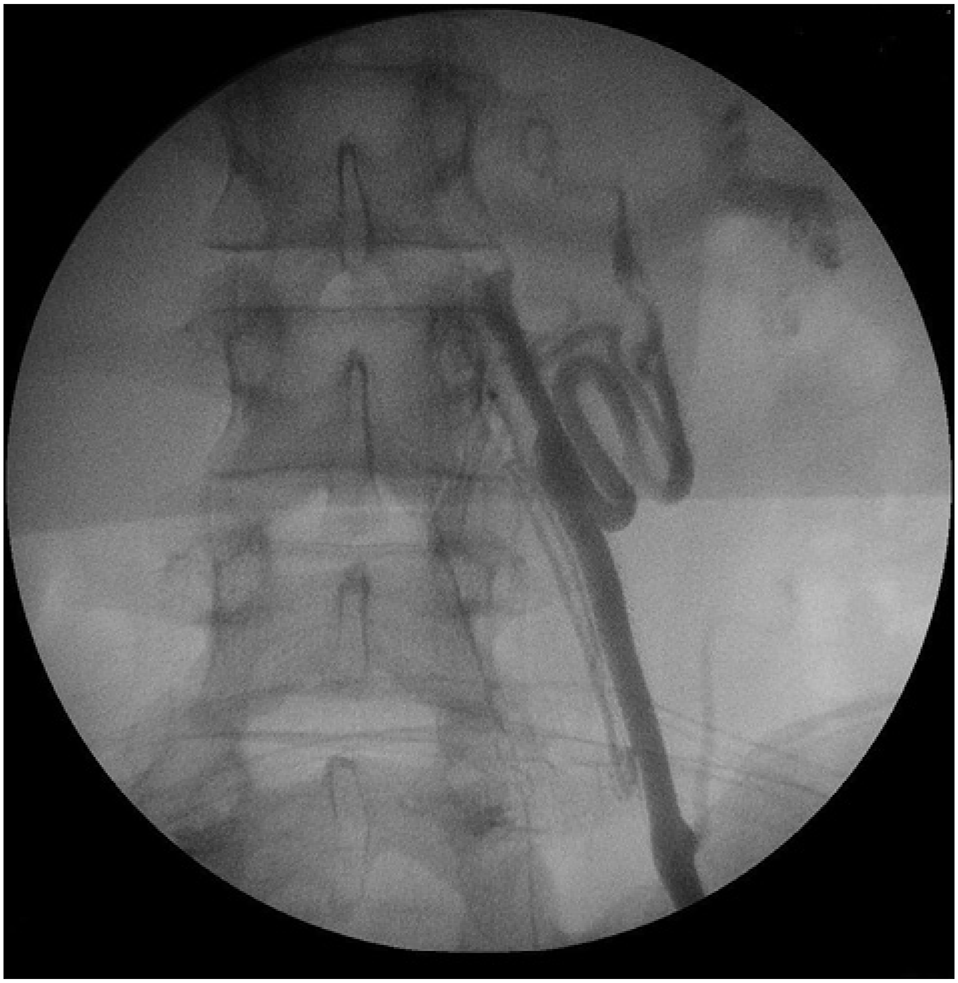
78.1a,b

A yellow 24-gauge cannula is inserted into the vein in a cranial direction and seen to bleed back before flushing with normal saline (**Figure 78.2**). The cannula is secured, and the screening C-arm is placed. The venous anatomy is confirmed by instilling Omnipaque® 300 mg/mL while screening continuously.



78.2

A microfoam is then prepared using a maximum of 2 mL Aethoxysklerol□ 3% and 8 mL air, mixed 20 times using a three-way tap. The patient is placed at 30 degrees head-up and a Valsalva maneuver is performed by the anesthetist. The microfoam sclerosant is then instilled into the vein under continuous screening injection and is terminated prior to reaching the renal vein (**Figure 78.3**). The cannula is withdrawn and the vein tied to prevent leakage of sclerosant. The spermatic fascia and wound is closed in layers with Vicryl□ sutures and skin glue.



78.3

OUTCOME

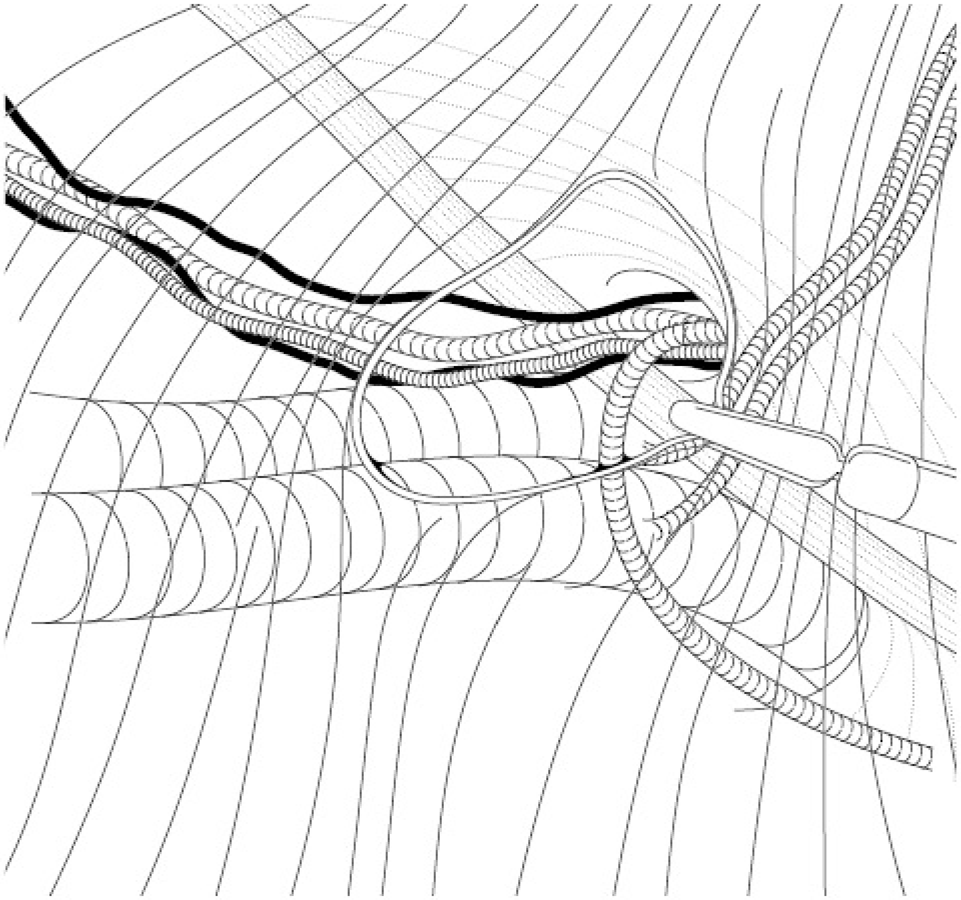
A prospective study of 91 patients undergoing antegrade sclerotherapy in our center has shown the success rate for the above procedure using microfoam to be 96%. The study compared liquid scleroscant via y-connector, liquid via cannula, and foam via cannula (success rates 58%, 90%, and 96% respectively). There were 11 recurrences across the three groups. Any persistent varicoceles were successfully managed with repeat sclerotherapy, and there were no instances of postoperative testicular atrophy. One postoperative hydrocele was managed surgically with a Jaboulay procedure.

Laparoscopic pediatric selective varicocelectomy (laparoscopic Ivanissevich procedure)

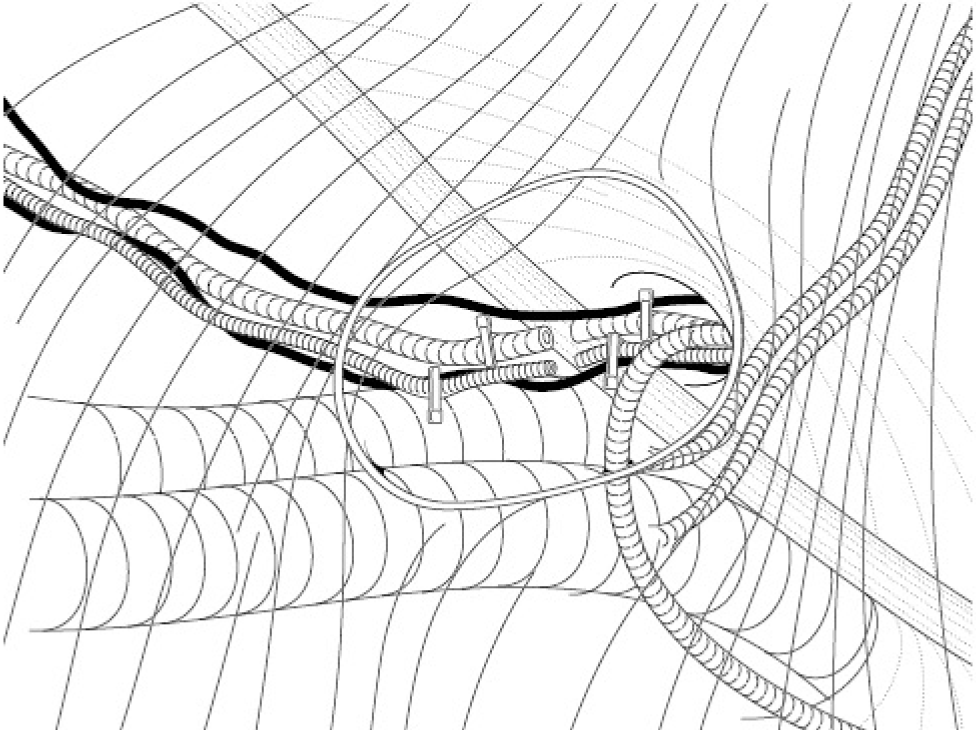
There is some confusion around the nomenclature of “varicocelectomy:” Oscar Ivanissevich described an open suprainguinal testicular vein ligation; Palomo later described suprainguinal ligation of both artery and veins; Bernardi actually described pampiniform ligation within the inguinal canal. A laparoscopic version of the Ivanissevich procedure is described here.

A pampiniform plexus vein is cannulated using the same methods as described previously. A mixture of 4 mL 1% methylene blue and 40 mL 300 mg/mL Omnipaque□ (iohexol) is then injected, and anatomy is confirmed with fluoroscopic screening.

A pneumoperitoneum is created using the open Hassan technique with a 5 mm 0-degree camera. Two 5 mm working ports (right lower quadrant and left upper quadrant) are inserted under vision. The peritoneum is divided to expose the vascular bundle proximal to the deep ring (**Figure 78.4a**). The testicular vein(s) are either selectively ligated using 5 mm endoscopic clips (**Figure 78.4b**) or sealed using a bipolar vessel-sealing device such as Plasmakinetic□ or Ligasure□ (Valleylab, Boulder, CO, USA). Methylene blue venography is then repeated to ensure all venous collaterals have been identified and ligated, with the aim of sparing both artery and retroperitoneal lymphatics.



(a)



78.4a,b

OUTCOME

In the authors' experience with 24 adolescent boys (median age 15 years) and a median follow-up of 18 months, there were three recurrences (12.5%). There were no postoperative hydroceles or testicular atrophy.

ACKNOWLEDGMENTS

Figures 78.1a,b and 78.2 are reproduced with permission from Tauber R, Pfeiffer D. Surgical atlas: Varicocele: Antegrade scrotal sclerotherapy. *BJU Int* 2006; 98(6): 1333–44.

VIDEO

OPS_8e_video_78.1 Antegrade sclerotherapy for varicocele

FURTHER READING

Keene DJB, Cervellione RM. Antegrade sclerotherapy in adolescent varicocele patients. *J Pediatr Urol* 2017; 13: 305.

Keene DJ, Cervellione RM. Intravenous methylene blue venography during laparoscopic pediatric varicocelectomy. *J Pediatr Surg* 2014; 49: 308–311.

Keene DJB, Sajad Y, Rakoczy G, Cervellione RM. Testicular volume and semen parameters in patients aged 12 to 17 years with idiopathic varicocele. *J Pediatr Surg* 2012; 47: 383–5.

Macey MR, Owen RC, Ross SS, Coward RM. Best practice in the diagnosis and treatment of varicocele in children and adolescents. *Ther Adv Urol* 2018; 10: 273–82.

Nagappan P, Keene D, Ferarra F, et al. Antegrade venography identifies parallel duplications in the majority of adolescents with varicocele. *J Urol* 2015; 193: 286–90.

Tekgül S, Dogan HS, Hoebeke P, et al. European Association of Urology (EAU) Guidelines on Paediatric Urology, 2016. Available at: https://uroweb.org/wp-content/uploads/EAU-Guidelines-Paediatric-Urology-2016-Pocket-1.pdf [Accessed 20 June 2020.]

79

Bladder exstrophy and epispadias

PETER M. CUCKOW AND KEVIN CAO

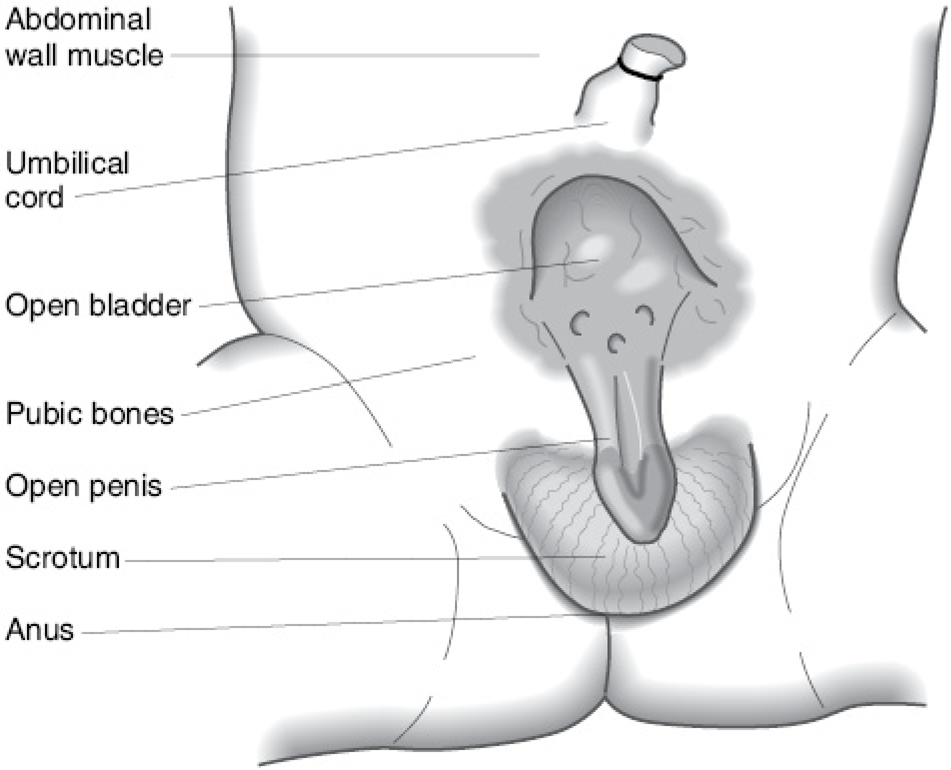
PRINCIPLES AND JUSTIFICATION

Contemporary estimates for the incidence of bladder exstrophy vary considerably. A recent international epidemiologic study placed the overall incidence at 2 per 100 000 births with a 2 : 1 male to female ratio. In the UK, the estimated incidence is 1 in 30 000 live births, with a 3 : 1 male preponderance. Primary epispadias has a similar male bias but is less common, with an incidence of 1 in 120 000. The surgical management of this rare group of anomalies is often concentrated in major surgical centres as in the UK, enabling the refinement of different surgical approaches.

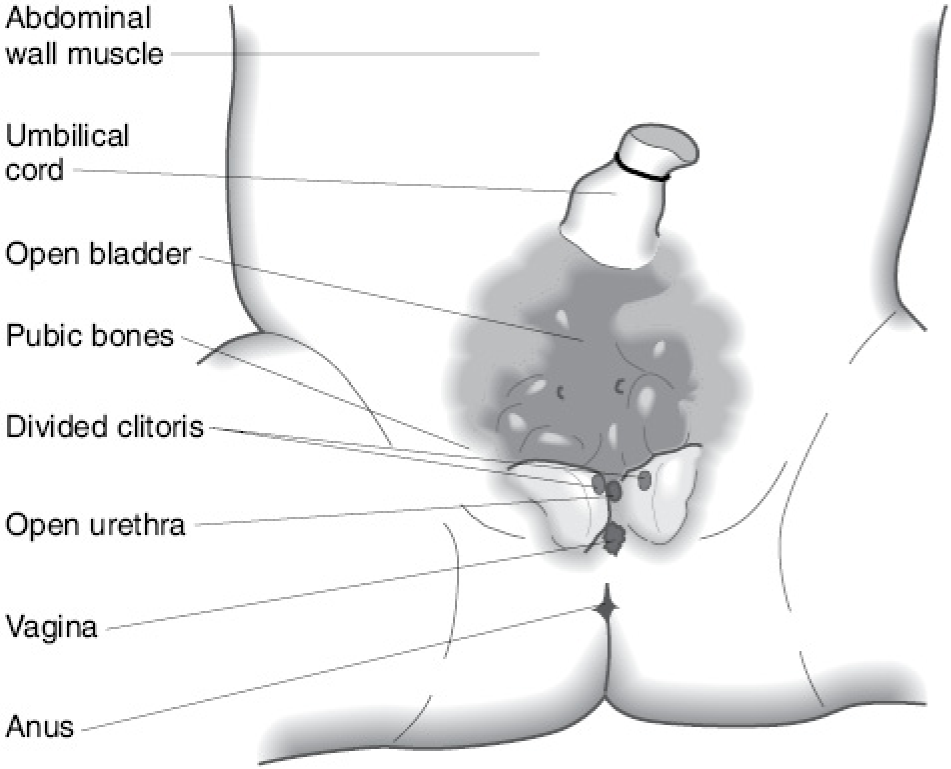
The aims of exstrophy and epispadias repair are:

- To place the bladder within the abdomen and ultimately to achieve urinary continence with preservation of renal function
- To create a satisfactory genital appearance and near-normal function
- To preserve or enable fertility

Bladder exstrophy presents at birth with a low-set umbilicus and a split rectus abdominis muscle attached on either side to separated pubic rami. The bladder occupies this low abdominal wall defect appearing as an open plate with the ureters draining directly onto its surface. It is continuous with the open urethral plate below, running down the dorsum of the penile corpora in the male and ending in an open glans (**Figure 79.1**). In the female, the urethral plate is shorter and runs between a divided clitoris and the labia minora to the anterior rim of the vaginal orifice (**Figure 79.2**). Inguinal hernias are a common association (~80%) and repair of bilateral herniotomies during the primary closure is commonly performed. Hernias are less common in girls (~15%) and so the need to perform routine herniotomy here is debatable. The perineum appears foreshortened, principally due to the incomplete development of the pubic rami and the anterior pelvic ring. The anus is anteriorly positioned, primarily because of this lack of structures in front, although its position may be appropriate relative to the structures behind, which are normally developed.

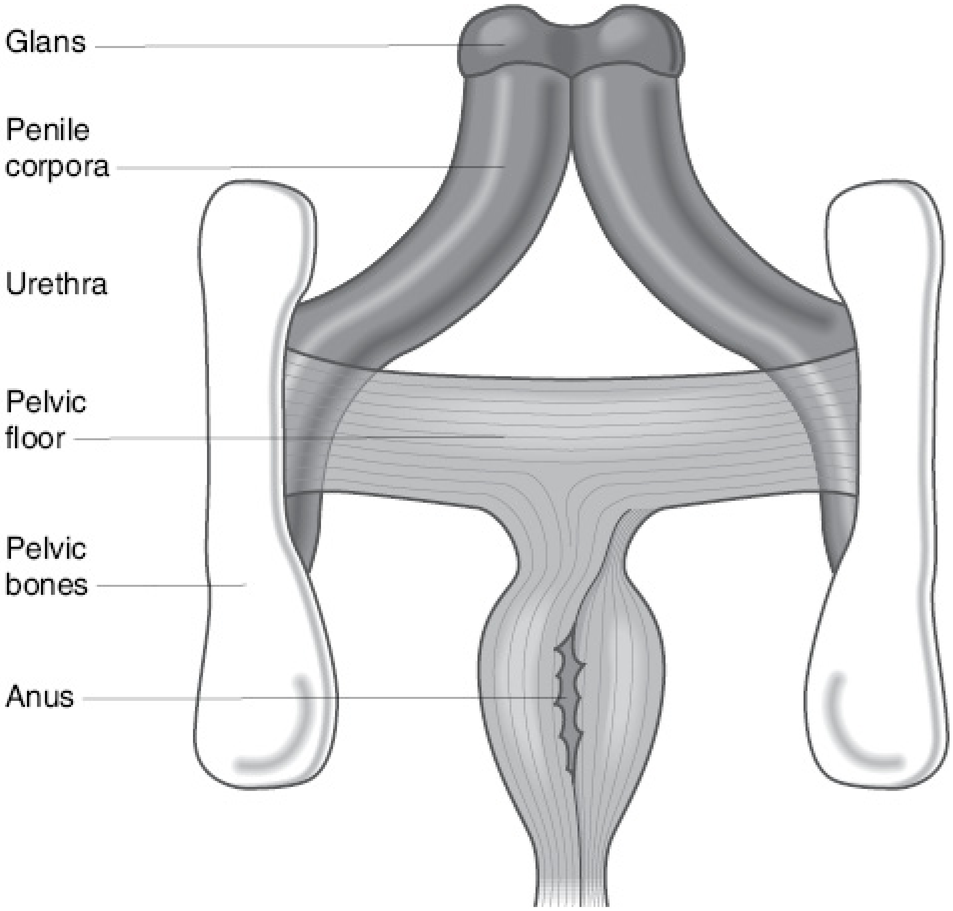

Abdominal wall muscle
Umbilical cord
Open bladder
Pubic bones
Open penis
Scrotum
Anus

79.1



79.2

Separation of the pubic bones in the midline (referred to as the diastasis) varies considerably. In primary epispadias, where only the urethra is open, the recti and the symphysis may be joined, whereas in the most severe forms of exstrophy the diastasis and separation of the rectus muscles at birth may be >5 cm. The size of the abdominal wall defect and the degree of difficulty in closure of the abdomen is directly related to the extent of the diastasis. The proximal ends of the penile corpora are attached on either side to the inferior pubic rami. While they may be shorter than normal corporal bodies, the separation of the pubic symphysis results in a Y-shaped penis and further limitation of the penile length (**Figure 79.3**). The urethra in the male is an open plate on the dorsum of the corpora and it has no relation to any sphincter muscles at birth. A normal sphincter complex is present in the perineum, however, which can be shown by direct stimulation in the area of the prostate gland between the proximal penile corporal attachments.

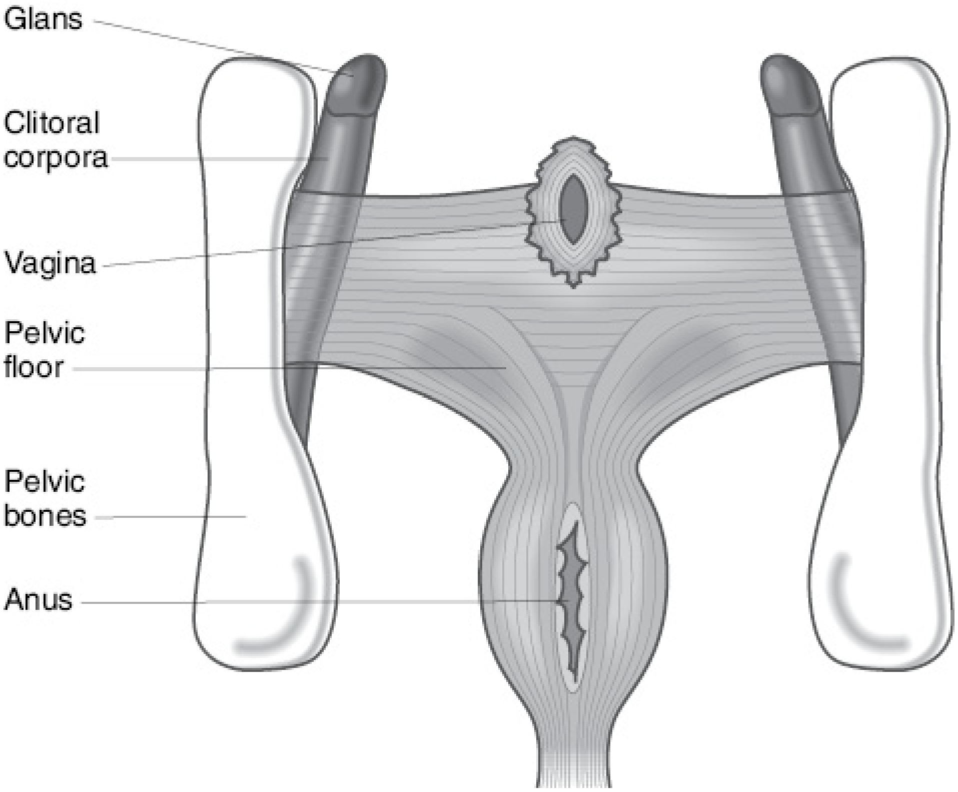


79.3

The inadequacy of the penis has led some in the past to opt for female sex of rearing for severe male cases. Current reconstructive procedures, however, with detachment of the penile corpora, now produce greater penile length. Sexual reassignment surgery is generally not performed in primary bladder exstrophy and cloacal exstrophy.

Isolated male epispadias varies considerably in severity, from that which simply involves the glans of the penis to a much more severe abnormality involving the entire urethra, bladder neck, and sphincter complex. In all but the least severe cases, patients have stress incontinence, and many bladders fail to achieve capacity.

There is a similar anatomical arrangement in females with primary bladder exstrophy. The divided clitoral corpora are attached to the pubic rami with muscle present, running between their origins (**Figure 79.4**). As the urethral plate is anterior to the vagina and not associated with this muscle complex, bladders fail to achieve capacity. The rare female isolated epispadias includes a bifid clitoris, a short patulous urethra, and an open bladder neck, invariably resulting in incontinence.



79.4

A reconstructive procedure is required for most cases to enable the bladder to fill, void, and achieve continence as well as to create a functional and cosmetically superior penis in boys. Our preference is the *Kelly soft-tissue reconstruction*, as described below.

This is preceded by closure of the exstrophy bladder shortly after birth, returning it to the lower abdomen and refashioning the abdominal wall. Without a bladder neck procedure, the bladder cannot adequately store urine and continence is only rarely achieved, with the child characteristically dribbling urine continuously. A successful primary closure, however, forms the basis for subsequent continence surgery. By performing a complete mobilization of the pelvic soft tissues, including the penile corpora, pelvic floor, and bladder, the bladder outlet, proximal urethra, and sphincter mechanism can be reconstructed. Variable bladder outlet resistance is a stimulus to its growth and an increase in capacity. These two factors combine to provide continence.

In addition, by mobilizing the penis in boys, greater penile protrusion is possible. In girls, a united clitoris and improved perineal cosmesis are achieved. In males, even greater penile length may be achieved by completely detaching the urethral plate from the dorsum of the corpora and creating a hypospadiac meatus. A subsequent urethroplasty is required to create the distal urethra and a terminal meatus. This procedure has been successful in cases where penile length is significantly impaired and may be a primary indication for the Kelly procedure in patients who have already had a continent urinary diversion. An alternative approach to the penis is to combine the soft-tissue mobilization with a *Cantwell–Ransley epispadias repair*. The decision to do this is made at the time of operation and depends on penile and urethral length.

Historically, patients have not normally become continent following exstrophy repair, requiring procedures that increase bladder neck resistance such as the *Young–Dees–Leadbetter reconstruction*, to achieve this. In many, an additional bladder augmentation cystoplasty (usually *ileocystoplasty*) and *appendicovesicostomy* (*Mitrofanoff channel*) are needed to create a large-capacity, catheterizable bladder. For a number of patients, this remains the ultimate management strategy, but the success of the Kelly procedure to enable per urethral voiding means that only around 30% of both boys and girls will still require augmentation surgery.

A lack of standardized assessment and nomenclature has led to much confusion about the results of treatment of bladder exstrophy. Our group has proposed a standardized definition of levels of incontinence in bladder exstrophy by which all interventions are assessed (**Table 79.1**).

Table 79.1 **Levels of continence in exstrophy**

Level	Continence
0	Dribbles urine constantly with no control
1	Able to retain urine with a “dry interval; ” some control, but still wearing protection
2	Sufficient dry intervals by day; in underwear and not needing protection; wet at night
3	Dry by day and night, no protection or accidents; “normal” as peers

SURGICAL STEPS TO CLOSURE

The surgical steps may be summarized as follows:

1. *Neonatal period:* Closure of the bladder without osteotomy or lower limb immobilization. In complex cases, restricted handling and continuous suction of the bladder urine with a Replogle tube postoperatively are advised. If the closure is delayed or in dehiscence cases, a Salter anterior iliac osteotomy with external fixation is recommended.
2. *Age 6–9 months:* Radical soft-tissue reconstruction (Kelly procedure). Bilateral ureteric reimplantation is usually performed at this time, to avoid compromise to the upper urinary tract that a combination of increased outlet resistance and ureteric reflux may bring.
3. *Age 2–3 years:* Completion of the urethroplasty in boys. Usually performed in two stages, with skin grafting to create a neourethral plate and glans groove on the penis, followed by urethral closure 12 months later.
4. *>Fifth year:* Bladder neck reconstruction, augmentation cystoplasty, and appendicovesicostomy are available for those in whom spontaneous voiding continence has not been achieved. Salvage surgery is usually not performed before the fifth year of life as continence may develop only slowly after the Kelly procedure.

Primary male epispadias is treated by the Kelly procedure if there is incontinence or poor penile length, or both. The timing of this procedure varies, but it may be performed after 6 months of age. Less severe cases (those that are continent) are treated by the Cantwell–Ransley epispadias repair.

Primary female epispadias is usually incontinent and so a Kelly soft-tissue mobilization is used.

POSTNATAL ASSESSMENT AND PREOPERATIVE EVALUATION

Many exstrophy cases are diagnosed *in utero* with characteristic combination of non-visualization of the bladder, low insertion of the umbilical cord, a short, thick phallus, and an irregular lower abdominal wall. Families should be counseled so that they understand the implications of the diagnosis and the postnatal surgical strategy.

Exstrophy closure should only be undertaken in well-resuscitated infants and may be delayed for up to 24–48 hours to allow the mother to recover from the delivery. Still, there is a persuasive argument to delay surgery further as time with the mother provides a better nutritional start and improved healing. Premature babies may require a longer period of medical stabilization before closure, which can be delayed for several weeks if this is medically indicated. Closure can be safely achieved without osteotomies up to 3 months of age in certain selected cases.

Renal ultrasound (US) should be performed to evaluate upper tract structure.

Oral clotrimazole suspension should be started to help prevent fungal colonization of the urinary tract. If osteotomy or lower-limb immobilization is planned, venous access to the lower limbs must be limited as these will be unavailable postoperatively.

A caudal anesthetic is helpful or, preferably, an epidural catheter is inserted to provide preoperative and postoperative pain control in both babies and older children having the Kelly procedure. In some difficult neonatal cases and where abdominal closure has been tight, a period of postoperative ventilation in an intensive care unit is required.

BLADDER EXSTROPHY CLOSURE

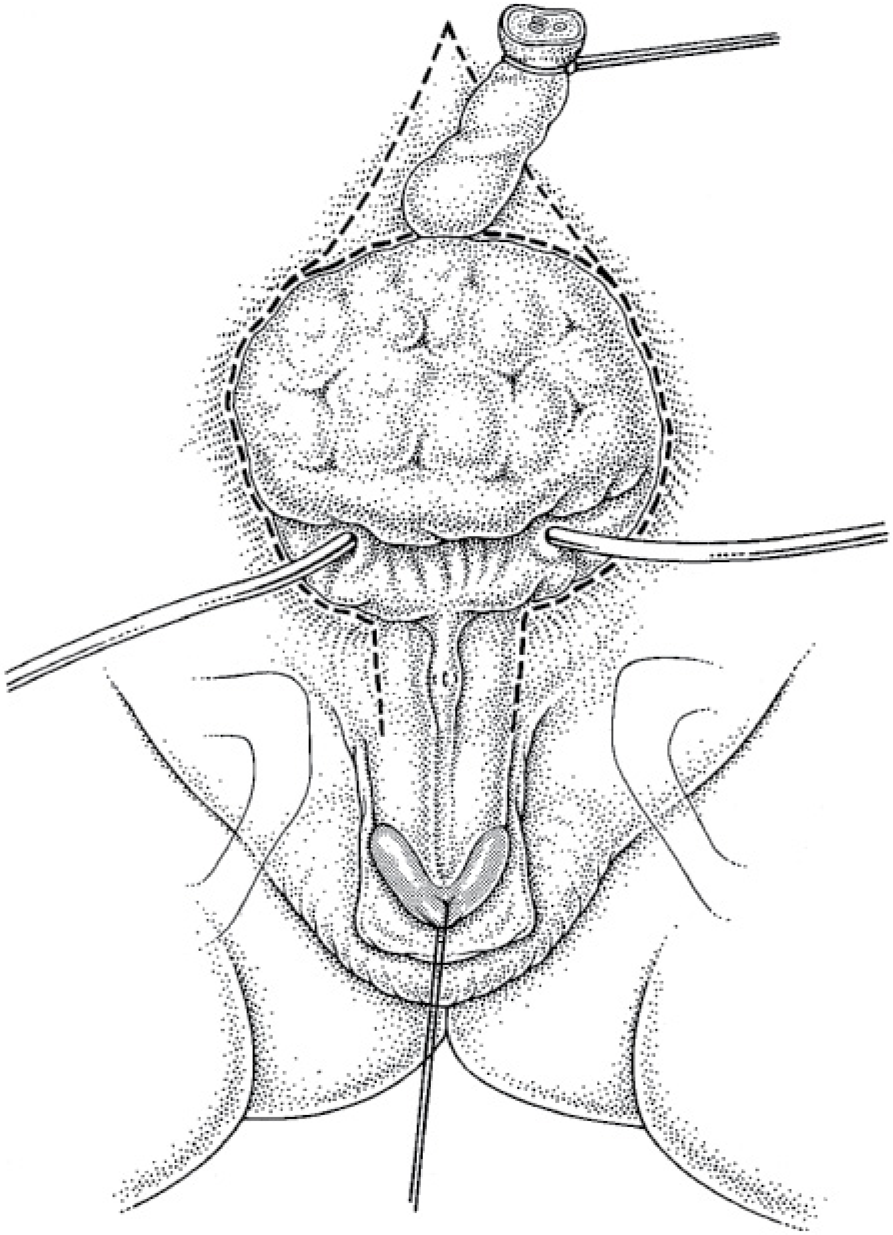
Operation

POSITION OF PATIENT

The patient is positioned flat and supine. Total lower body preparation with individual wrapping of the legs allows maximum mobility at the time of the surgery and access for the orthopedic surgeons to perform osteotomies if required. Amikacin (7.5 mg/kg), metronidazole (7.5 mg/kg), and ampicillin (6 mg/kg) are given intravenously in the anesthetic room and continued postoperatively for at least 48 hours.

SKIN INCISION

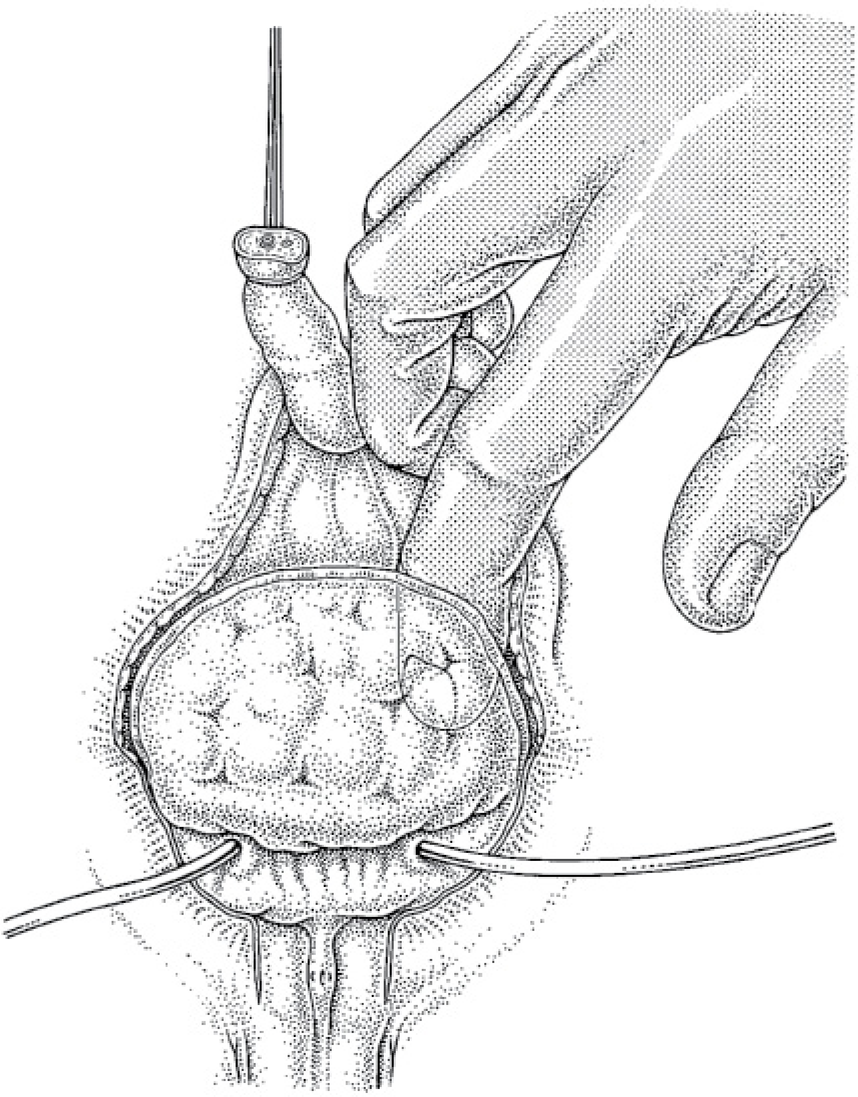
The umbilicus is ligated and trimmed but retained. A glans stay suture will aid retraction. Incisions are made beginning in the midline above the umbilicus, extending around the margins of the bladder, onto the root of the penis and on either side of the urethral plate as far as the distal limit of the verumontanum (**Figure 79.5**). The incisions are deepened with diathermy but the distal incisions may be left superficial at this stage.



79.5

BLADDER MOBILIZATION
The incision is deepened through the midline to expose the umbilical vein, which is ligated, without opening the peritoneum. The umbilical arteries serve as a guide on each side to the extraperitoneal plane. Both are divided, allowing the umbilical stump (which will become a source of wound

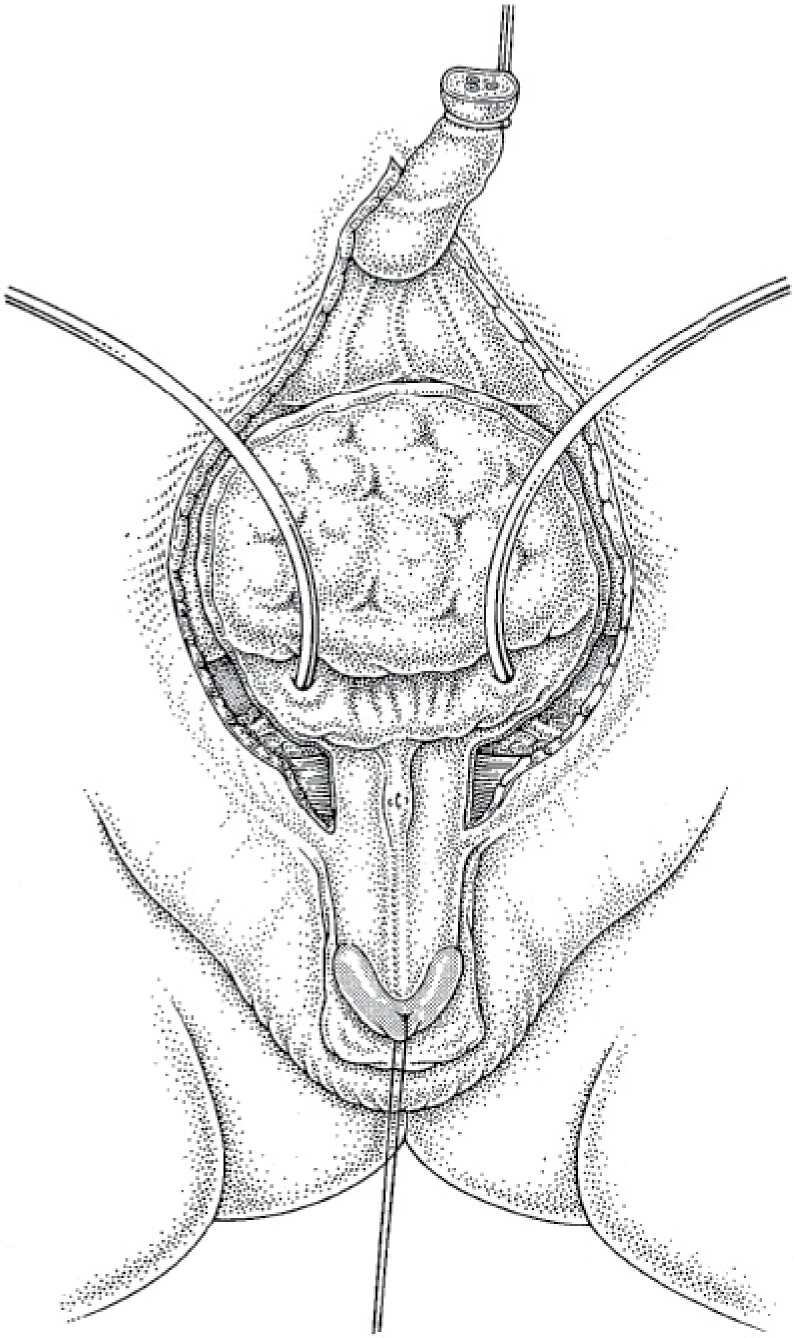
infection later) to be removed. Both ureteric orifices should be catheterized with 4 Fr or 6 Fr catheters and careful, blunt dissection will open a plane behind the rectus muscles on each side, in front of the ureters (stent in position for palpation), and down to the pelvic floor at the bladder neck (**Figure 79.6**).



79.6

COMPLETION OF DISTAL INCISION

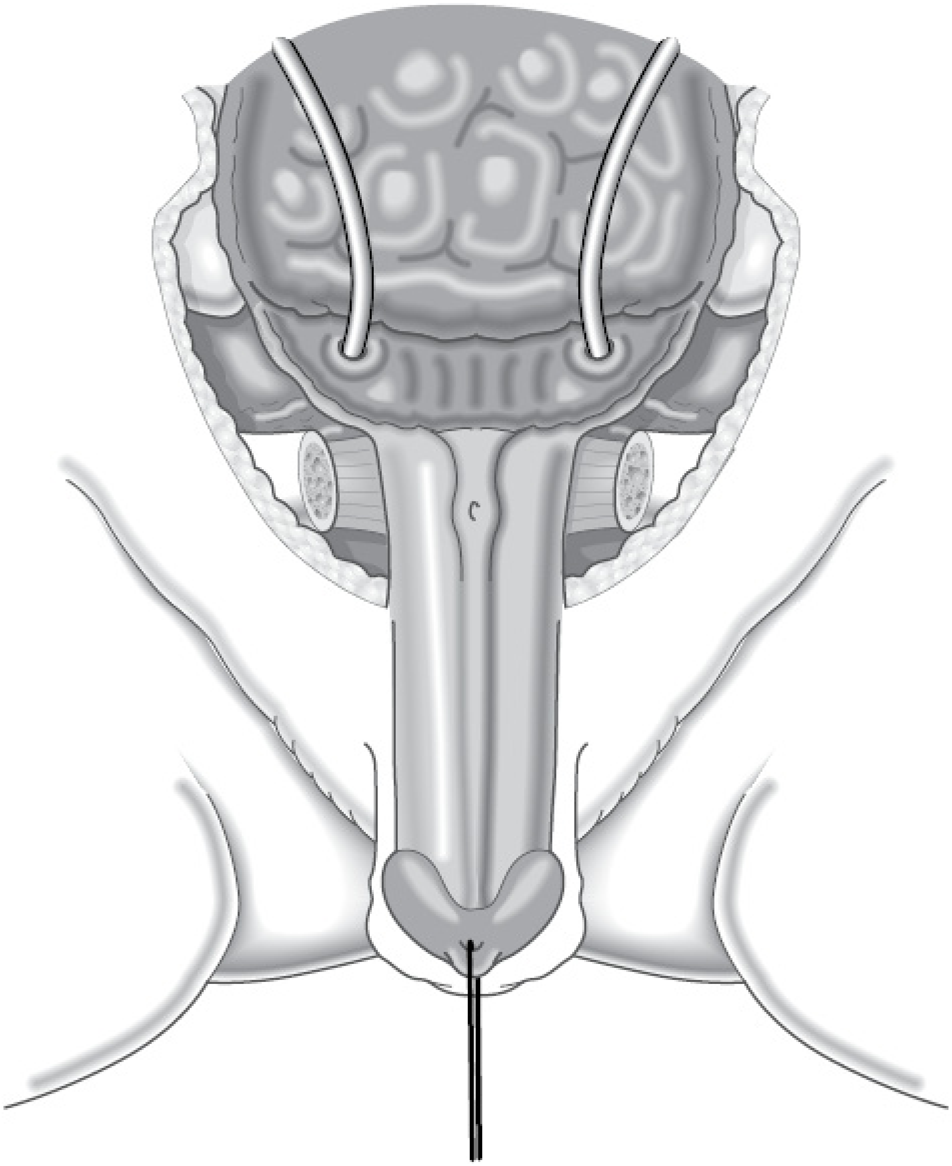
With a finger in position behind the abdominal wall, the distal incisions may be completed with diathermy to create freedom of the bladder on each side. The intrapubic bar tissue fusing with the bladder neck is now visible and is an important landmark (**Figure 79.7**).



79.7

DISSECTION OF PROXIMAL URETHRA

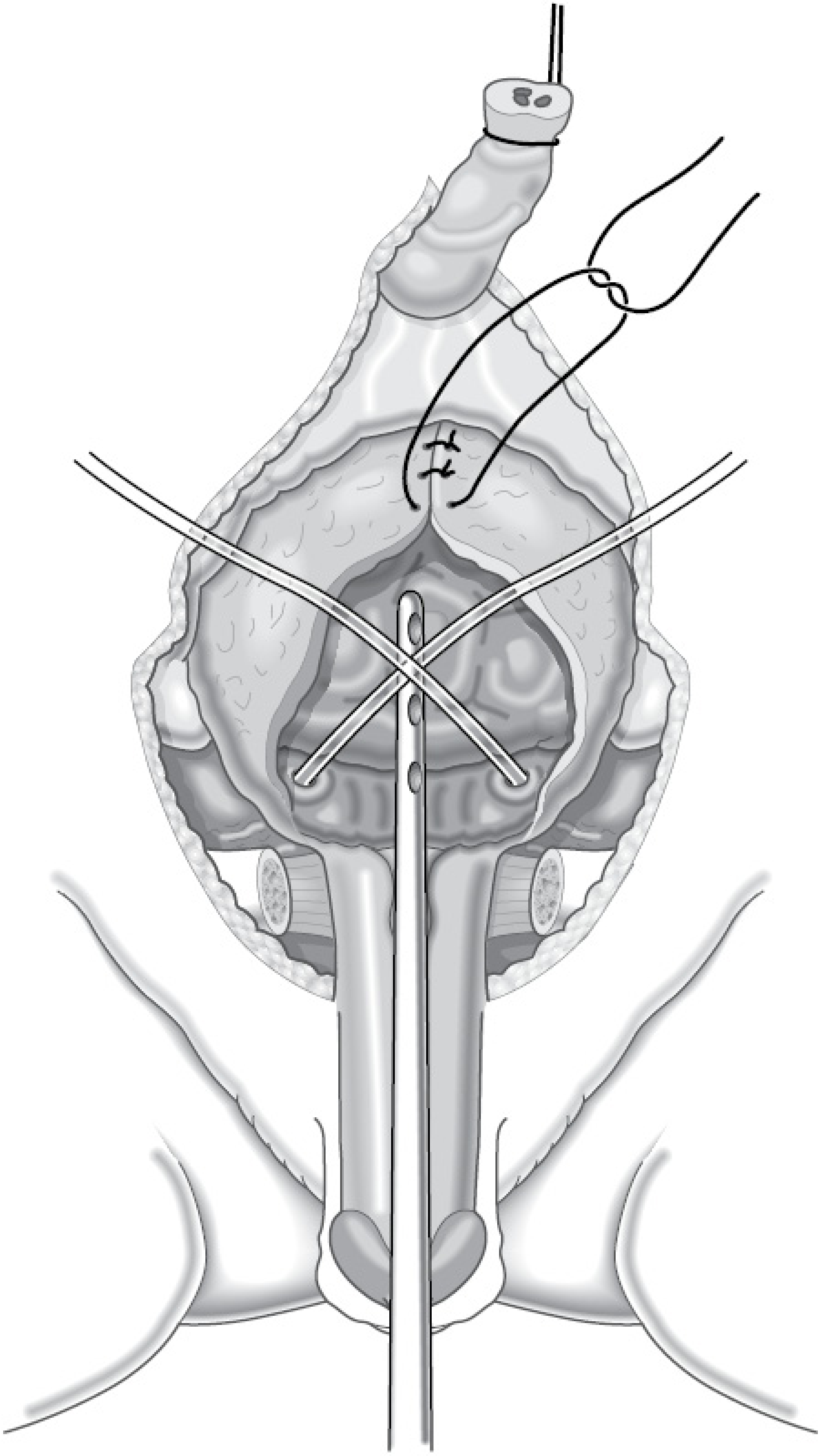
The intrapubic bar tissue is divided at the lower end of the incisions, lateral to the bladder neck area, above the verumontanum, to allow the bladder's placement inside the pelvis (**Figure 79.8**). This allows the edges of the wound and the pubic bones within this soft tissue to be approximated in front of the bladder neck. Radical corporal mobilization is not performed at this stage, although incisions in the intrapubic tissue may need to be extended distally to facilitate the closure of the pelvic ring. It is preferable to stay above the corporal bodies in patients subsequently having a Kelly procedure.



79.8

BLADDER CLOSURE

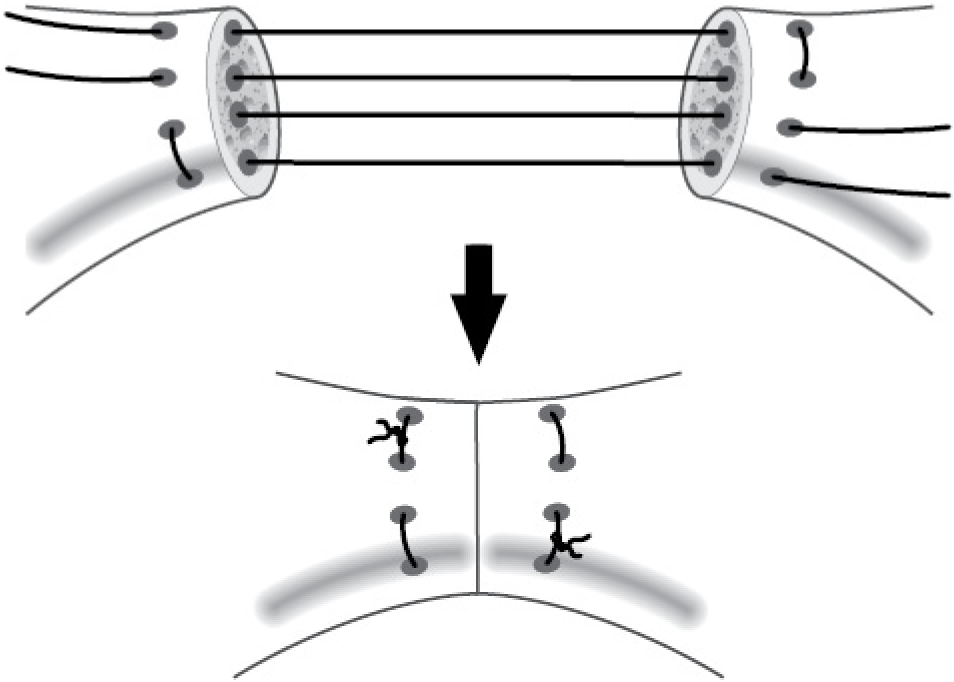
The bladder and proximal urethra are now sufficiently free to drop back into the pelvis. An 8 Fr feeding catheter with opposed eyes to avoid obstruction is sutured to the bladder wall with 6/0 Monocryl™. Bladder closure begins from the apex using interrupted 4/0 Monocryl™ sutures after trimming the bladder edges. The two ureteric feeding tubes (which have also been secured with 6/0 Monocryl™ sutures) are brought out between sutures. Closure is continued to the proximal urethra, although the lower sutures may be left untied until pelvic approximation is achieved (**Figure 79.9**).



79.9

APPROXIMATION OF PUBIC BONES

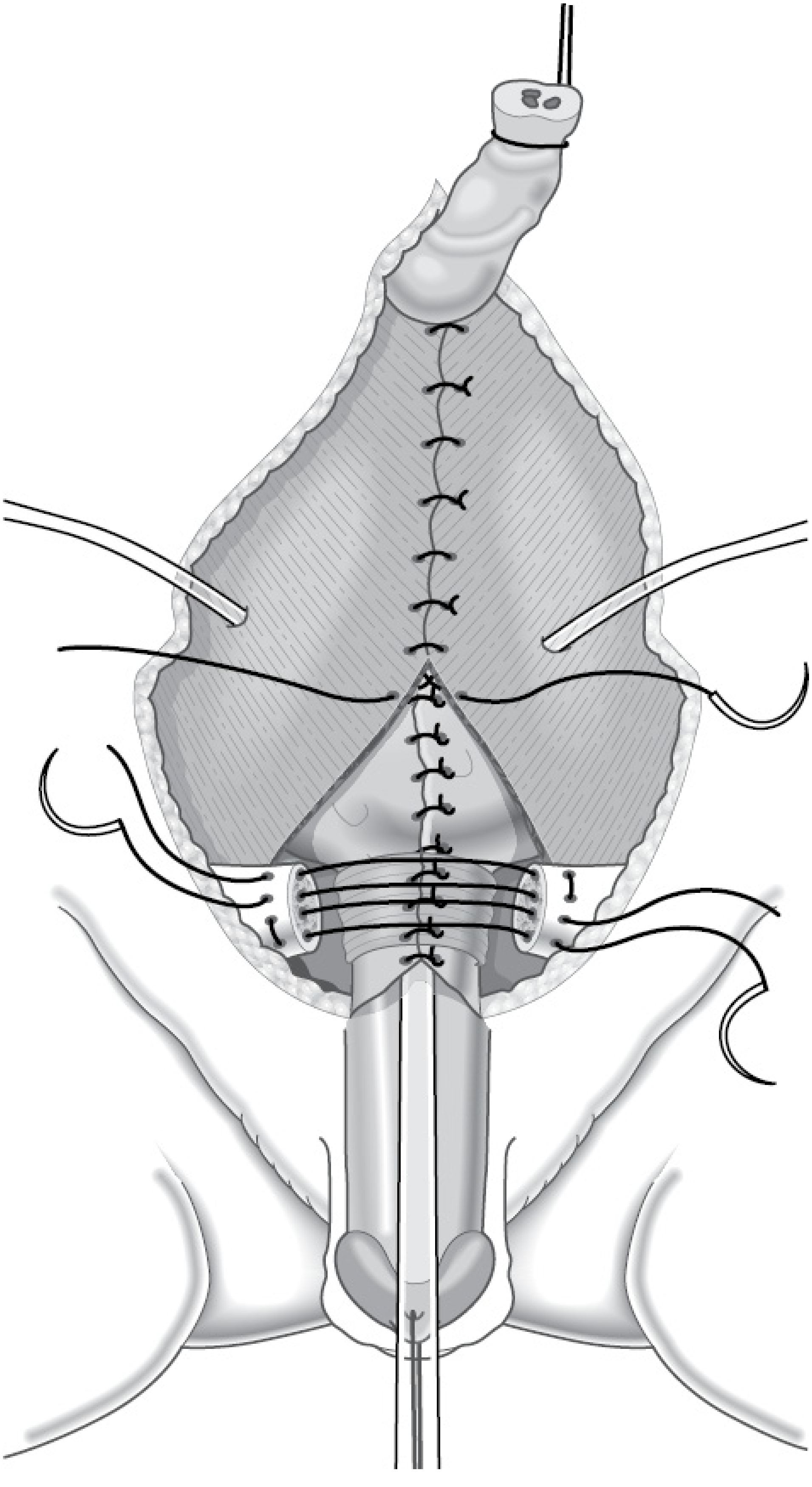
Two mattress (0 or 1) polydioxanone sutures (PDS™) are placed between the pubic bones and run within the soft tissue to the medial edge of the lower part of the wound. This will allow approximation of the lower wound in front of the bladder without there being a potential bowstring of suture material to cut through the structures behind (i.e. the bladder neck and urethra). The abdomen is closed from above, using figure-of-eight sutures to bring the rectus muscle edges together. This is completed by tying the pubic sutures at the end (**Figure 79.10**).



79.10

DEEP AND SUPERFICIAL WOUND CLOSURE

The umbilicus has usually been excised. The ureteric catheters are tunneled lateral to the rectus, using a cannula. A further layer of fat sutures of 4/0 Monocryl™ are used to cover the deep closure before a row of interrupted or a continuous suture closes the skin edges (**Figure 79.11**). A dressing of skin glue further takes some tension out of the wound and provides a temporary waterproof covering above the urethral opening.

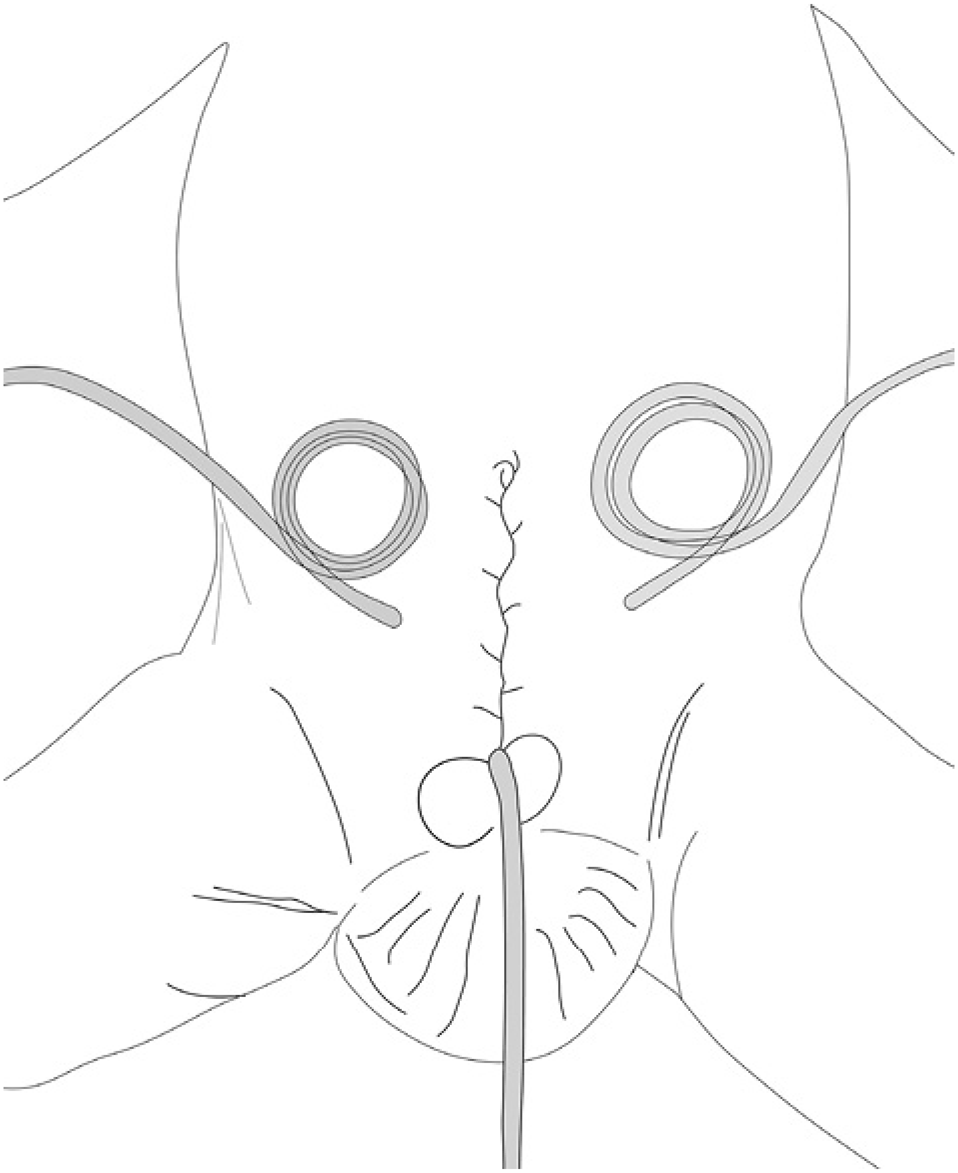


79.11

This is done throughout the whole length of the wound so that the strain of closure is not taken by the symphysial sutures alone. Interrupted figure-of-eight sutures of 3/0 PDS™ are used proximally, increasing to 2/0 PDS™ as the closure proceeds distally. The ureteric stents are brought out on either side of the abdomen by cutting off their connectors and threading them onto large-bore IV cannulas, introduced from the outside. Finally, the pubic bones are approximated by tying their sutures. During this maneuver, internal rotation of the hips by the assistant and compression of the pelvis may be helpful.

Postoperative care and immobilization

We do not recommend any form of pelvic immobilization, noting in our patient cohorts that successful closure is achieved in more than 95% without it. This enables normal parental care immediately after surgery and a reduction in length of stay and medical costs by abandoning osteotomies and pelvic traction/casting. Ureteric stents should be maintained for at least one week to keep the bladder closure dry during healing. The urethral catheter should be left for a little longer or until it falls out spontaneously (**Figure 79.12**). Ultrasound is undertaken at 6 weeks post-surgery and the child is maintained on prophylactic antibiotics.



79.12

RADICAL SOFT-TISSUE RECONSTRUCTION (KELLY PROCEDURE)

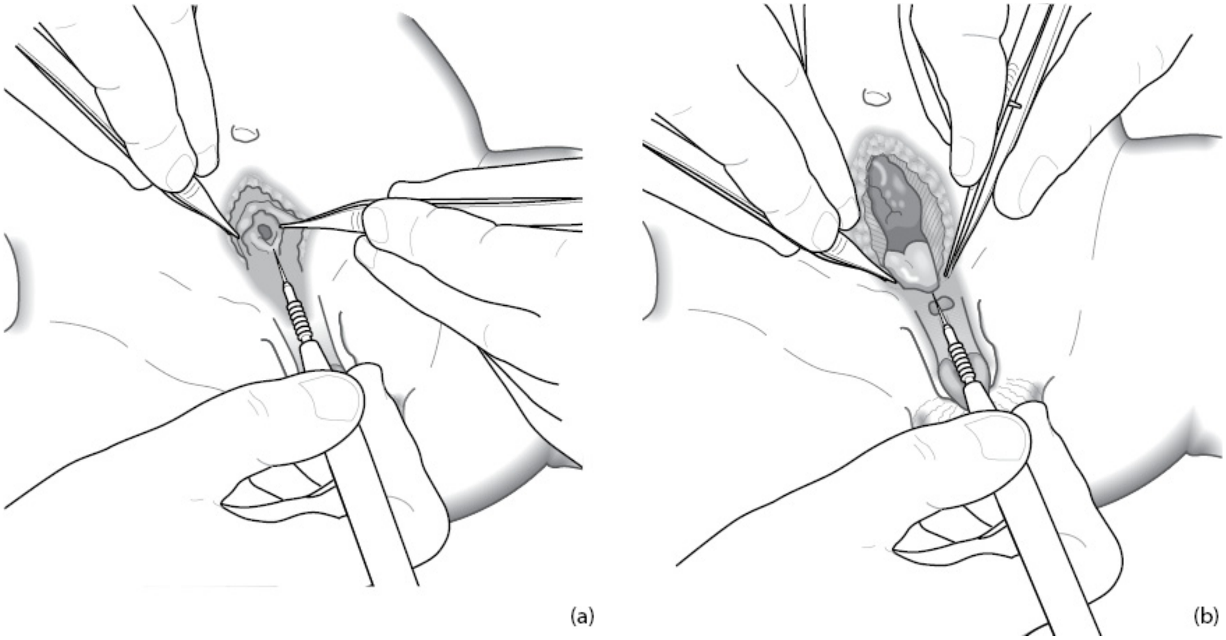
This operation reopens and dissects the bladder, mobilizes the urethral plate completely from the penile corpora, releases the pelvic floor muscles, and detaches the penis from the pubic rami, preserving its neurovascular bundle (the pudendal pedicle). It allows radical reconstruction of the bladder neck and urethra as well as recreating a muscular sphincter. A major benefit is the improvement in penile length that is achieved.

There are then two options in respect of the penile reconstruction. The first is to combine the operation with a Cantwell–Ransley epispadias repair, which leaves the urethral plate attached distally to the glans and enables a single-stage procedure but at the expense of some penile length. Alternatively, the urethral plate can be completely detached, which frees the penis for more length but leaves a hypospadiac meatus, requiring a completion distal urethroplasty.

Operation

SKIN INCISION

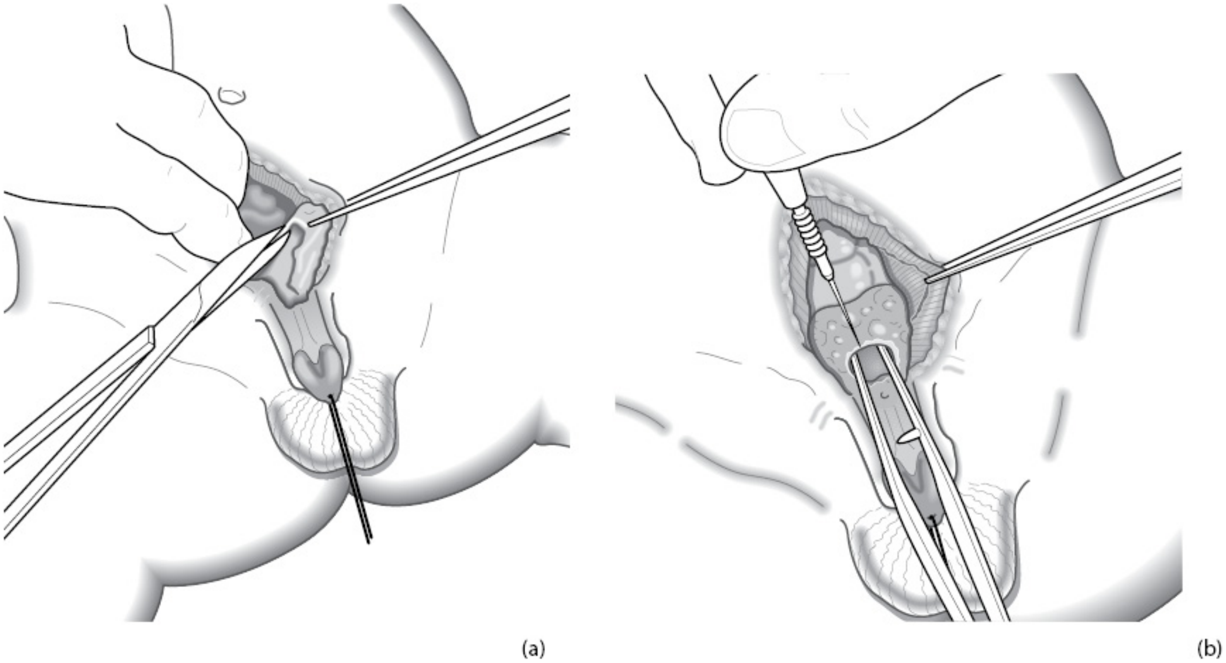
A glans stay suture is placed for retraction. The old incision is reopened, using an incision that will make a strip of midline skin and the scar from the previous closure available for the creation of a new umbilicus. This is deepened through the muscle to expose the bladder below and the peritoneum above (**Figure 79.13a**). The bladder is carefully dissected from behind the abdominal wall, taking care to preserve the detrusor muscle. The bladder is opened in the midline to expose the trigone and the ureteric orifices but limited in length superiorly so a hand retractor can be inserted to gain better exposure of the trigone (**Figure 79.13b**).



79.13a,b

BLADDER MOBILIZATION

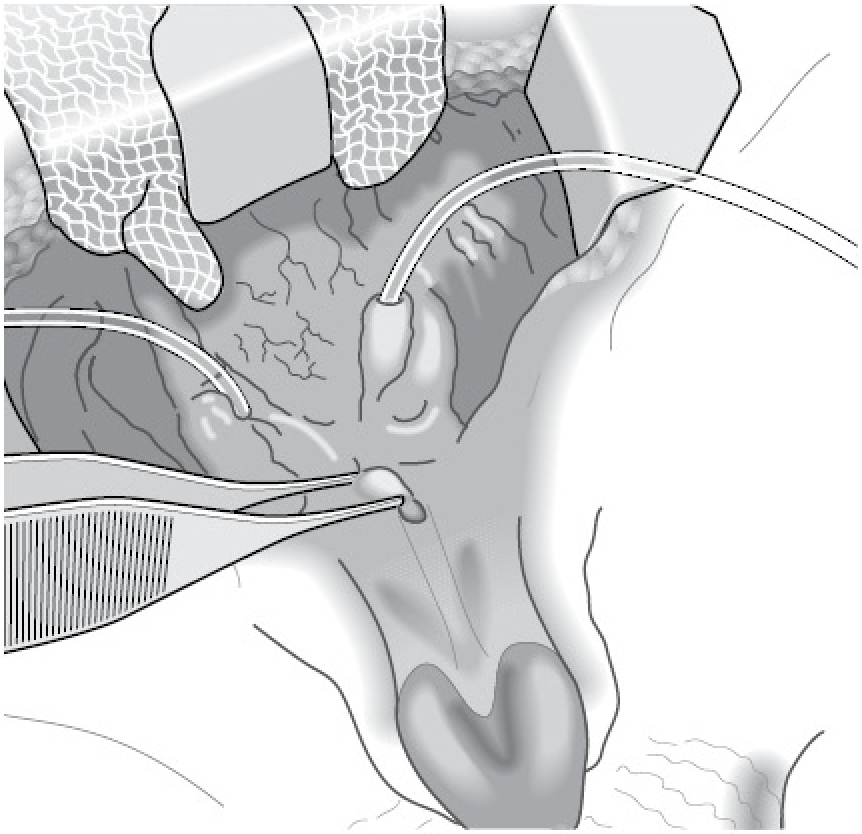
The lateral edges of the bladder are freed from the abdominal wall and the bladder is reopened anteriorly in the midline (**Figure 79.14a**). The lateral edges of the bladder are dissected free to the level of the verumontanum, as in primary repair (**Figure 79.14b**).



79.14a,b

URETERIC REIMPLANTATION

Both ureteric orifices should be catheterized with 6 Fr feeding tubes. Keeping close to them, both ureters should be dissected and a Cohen ureteric reimplantation completed with a transverse tunnel (**Figure 79.15a–c**).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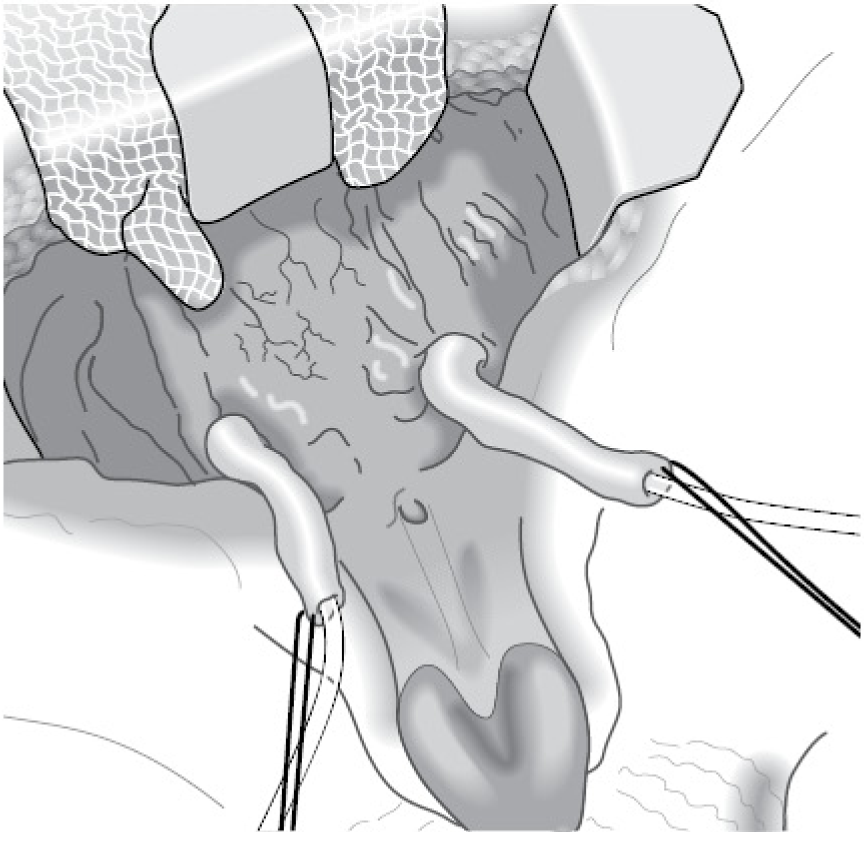

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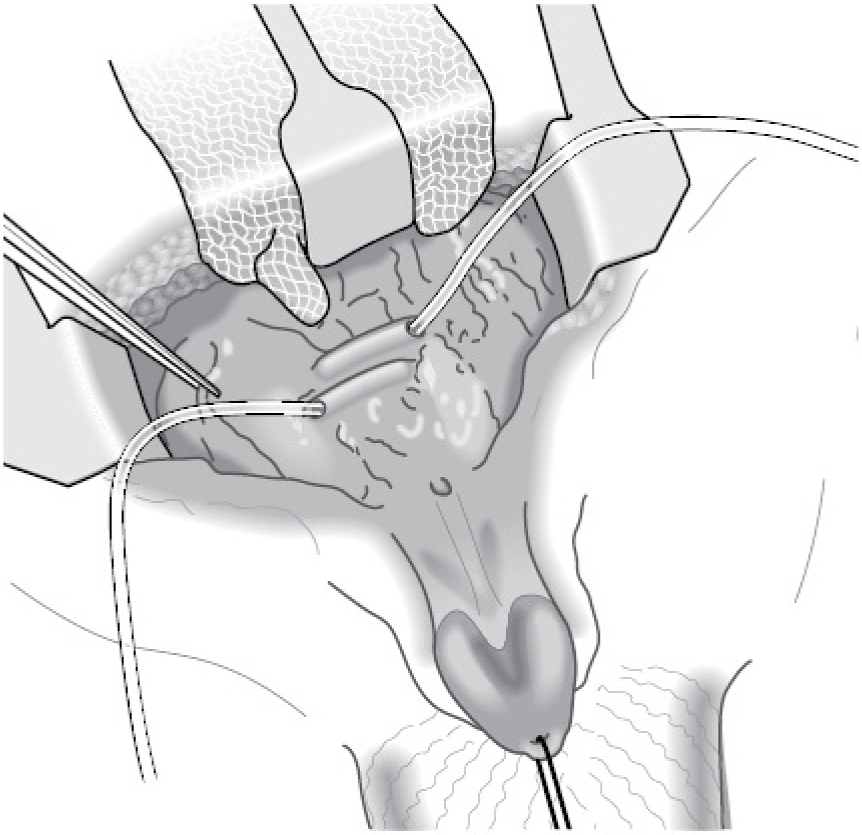
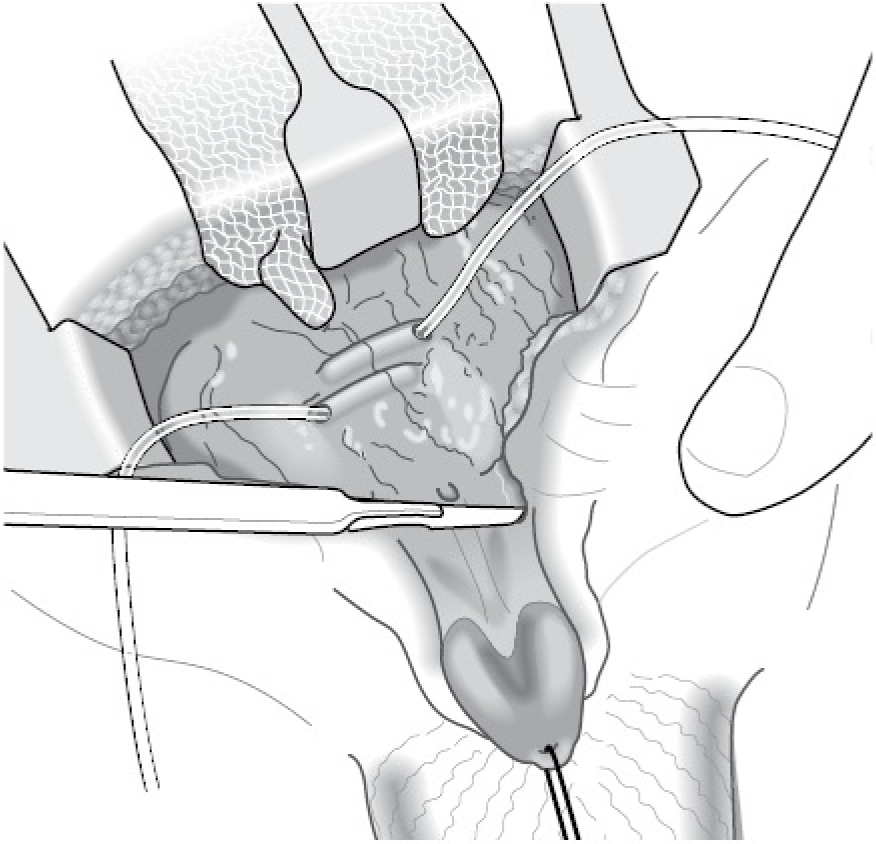


(c)

79.15a–c

URETHRAL AND PENILE SKIN DISSECTION

The urethral plate is marked out from either side of the verumontanum, distally to the edge of the corpora, and incised. Ventrally, the penile skin is marked and incised to leave a cuff beneath the glans. The skin is lifted off the ventrum and sides of the penis and dissected proximally to expose the undersurface of the corporal bodies (**Figure 79.16a–c**).



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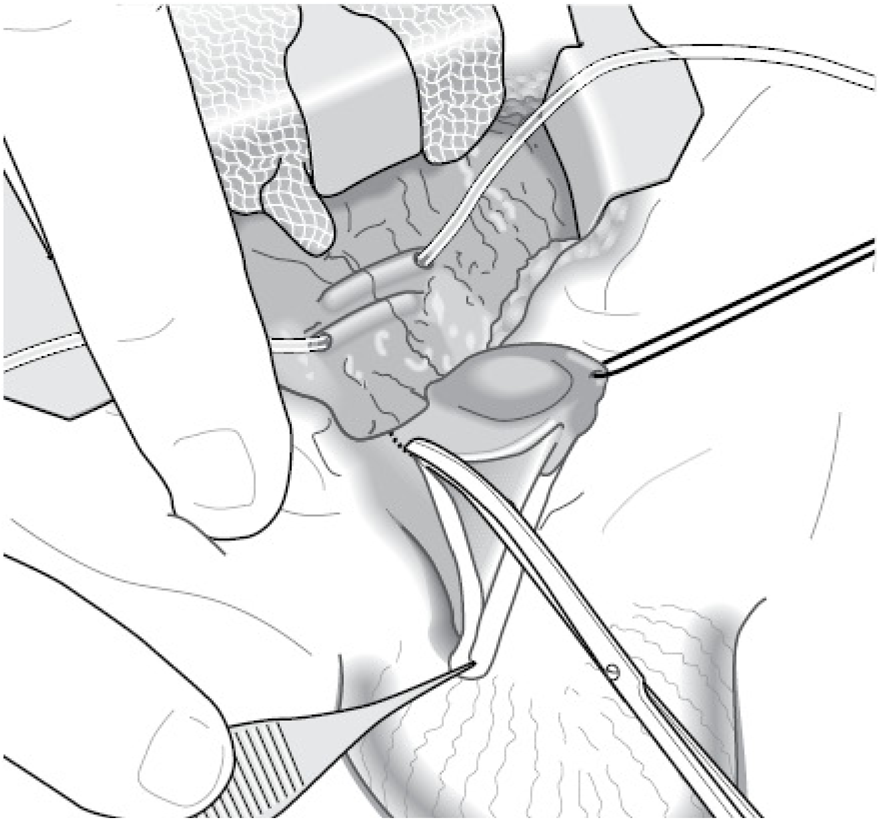
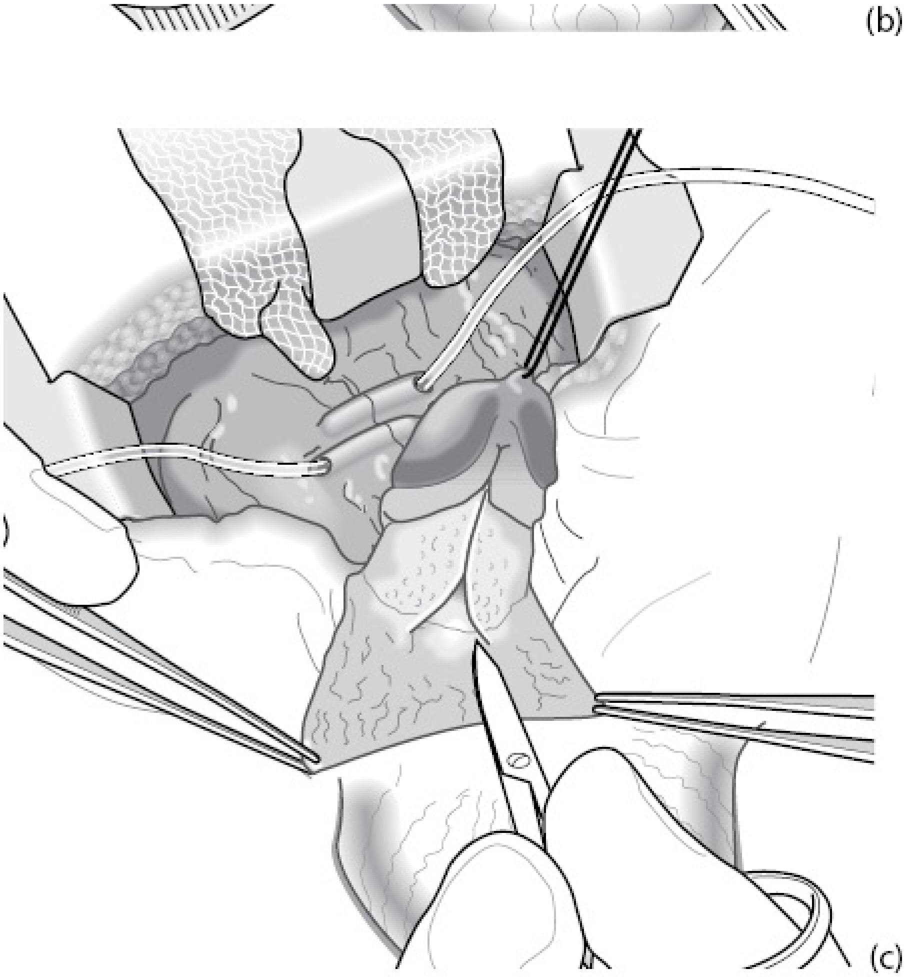
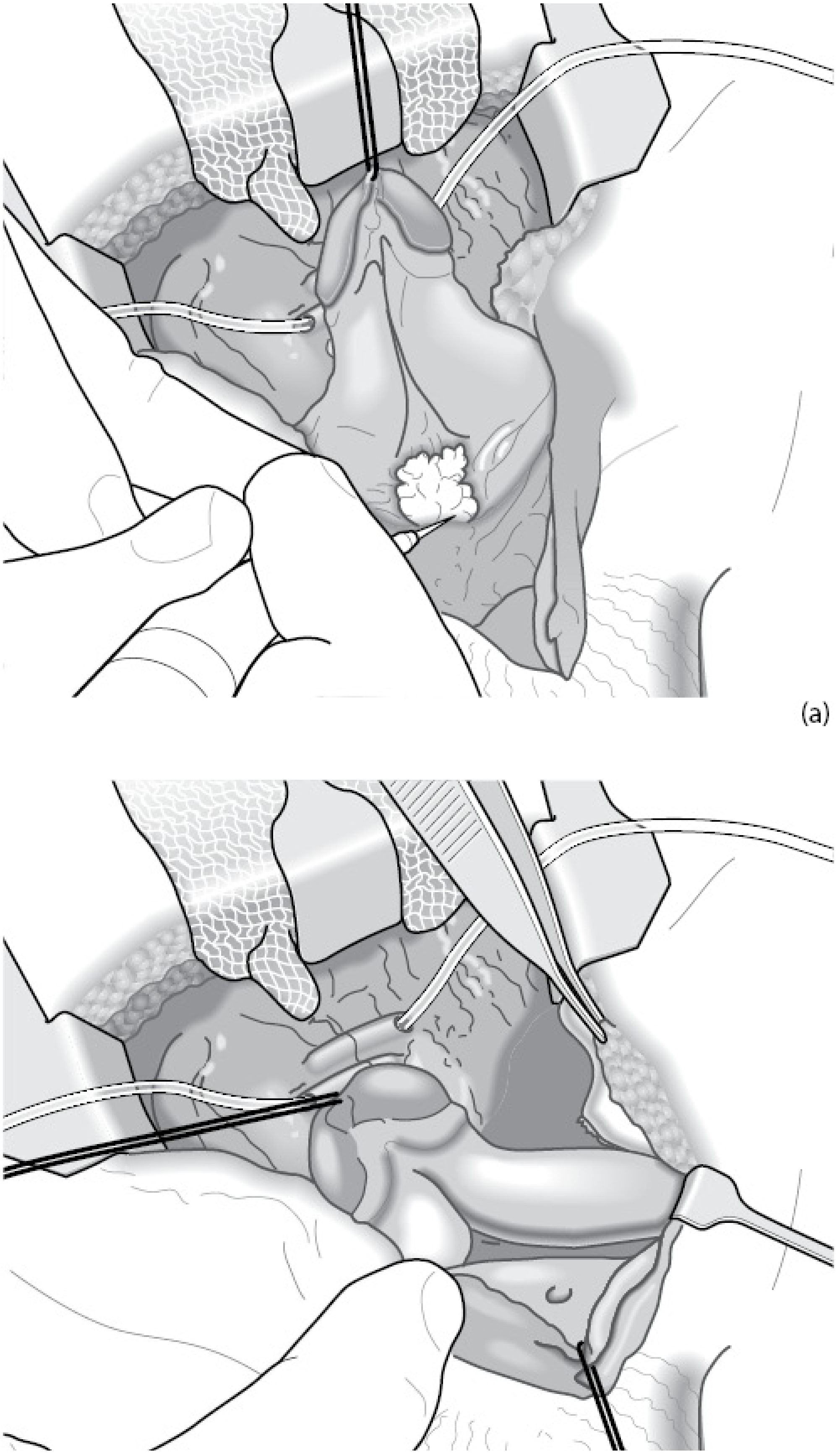




79.16a–c

DISSECTING THE PENIS

Dissection continues proximally along the corporal bodies. The separation of the corpora and the prostate gland in between them are identified. Laterally, each corpus is traced until its point of attachment to the inferior pubic ramus can be seen and palpated. The perineal muscle can be identified with the muscle stimulator, running between the bases of the corpora (**Figure 79.17a–c**).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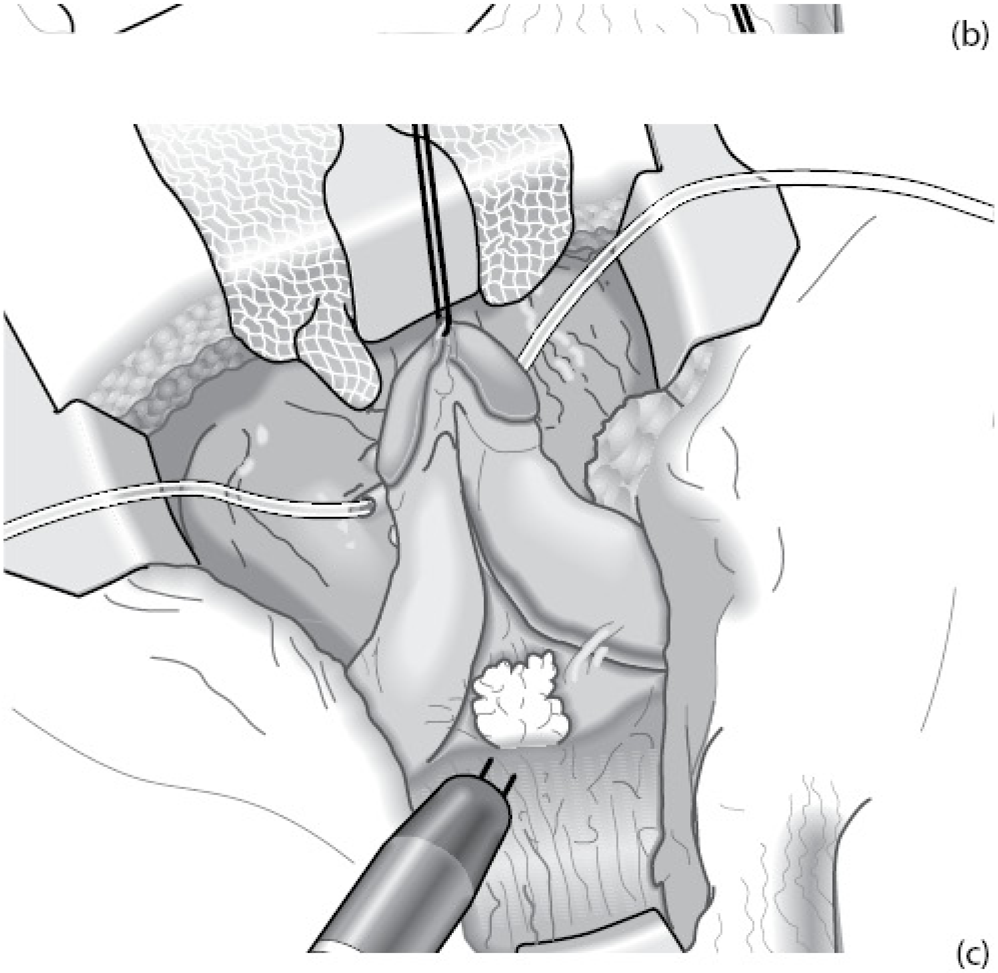
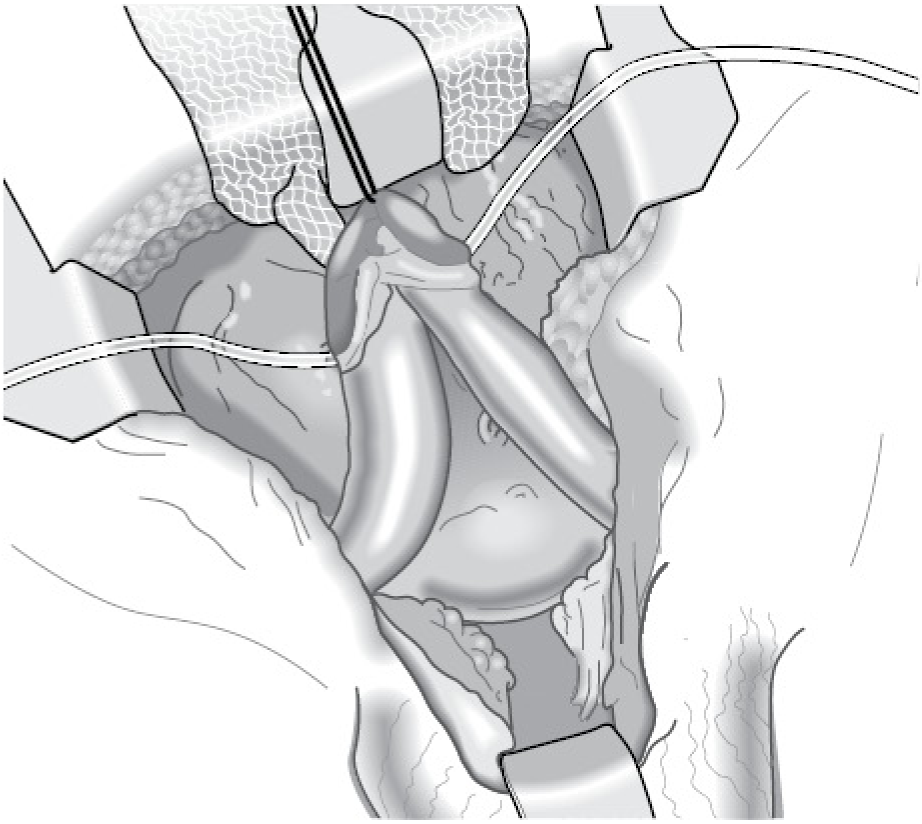


79.17a–c

DISSECTING THE URETHRAL PLATE

The urethral plate is dissected from above and below the penis. The tissue below the plate is separated from the corporal bodies by dissecting the bloodless plane between them on both sides, keeping close to the corpora. Dissection is completed anteriorly on either side of the plate, and vessel loops are passed around each corpus. Separation of the plate is completed, distally to the edge of the glans and proximally to the level of the prostate (**Figure 79.18a–c**).



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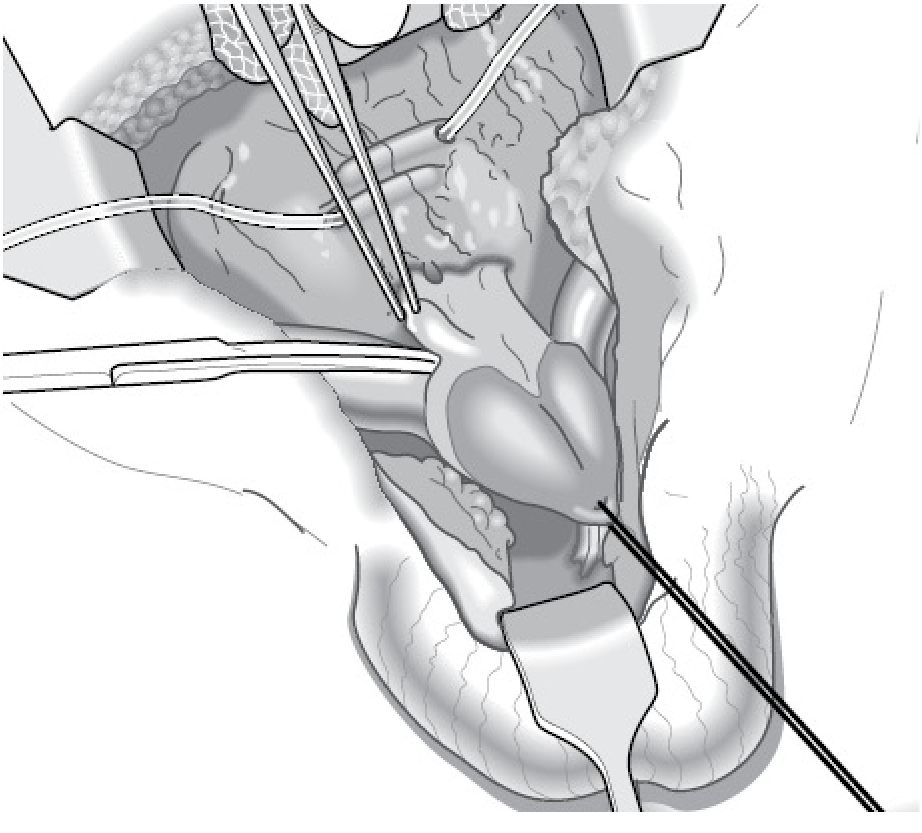
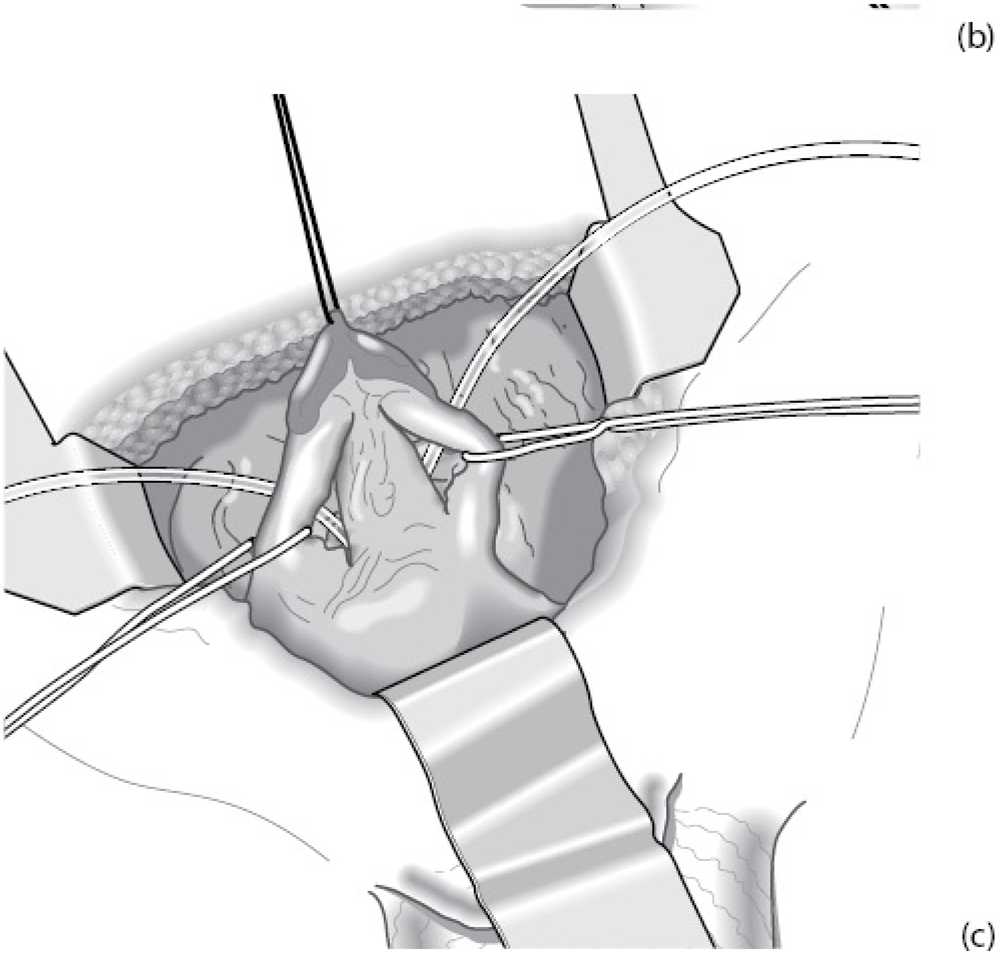
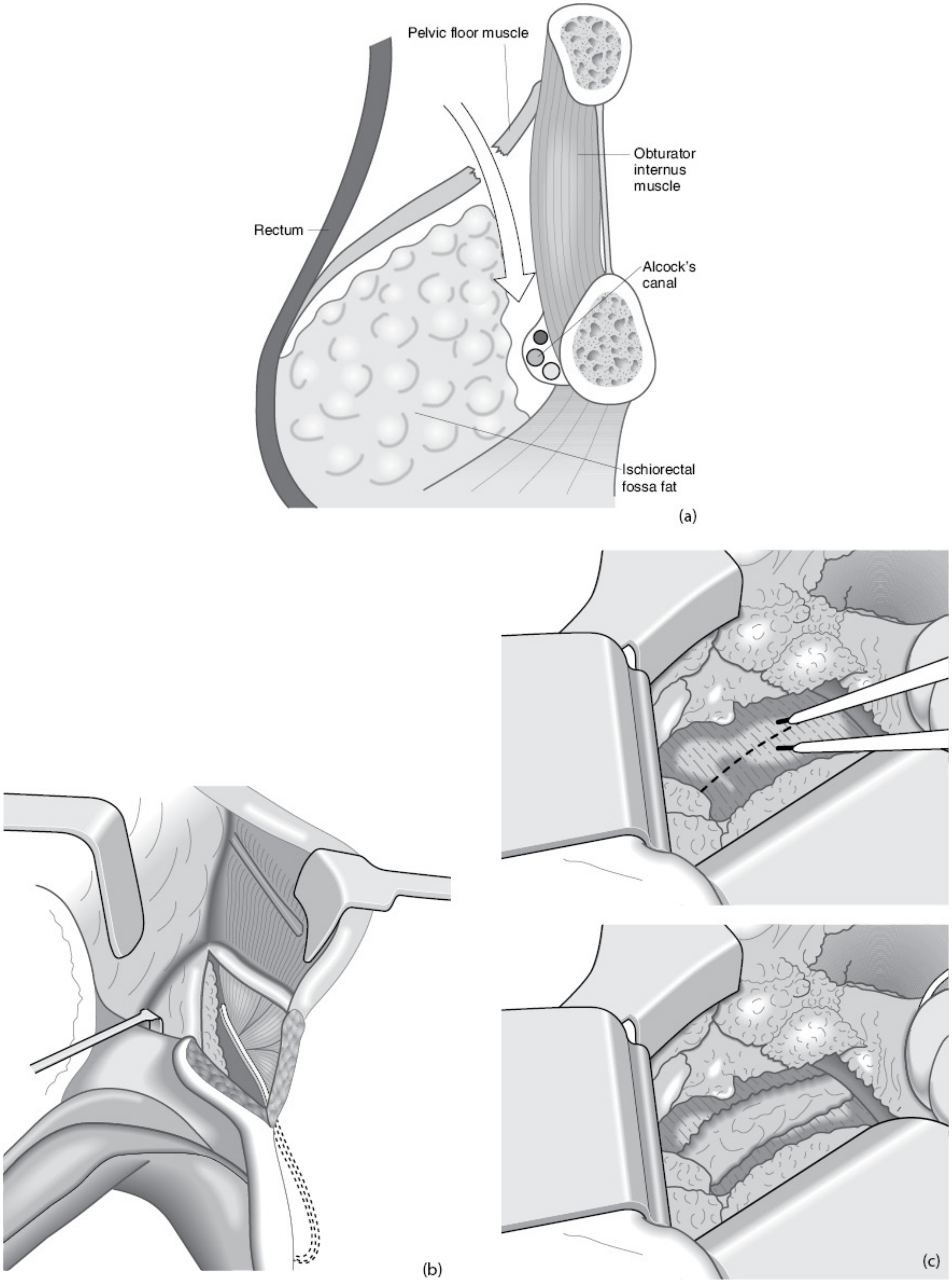




79.18a–c

RELEASING THE PELVIC FLOOR FROM ABOVE

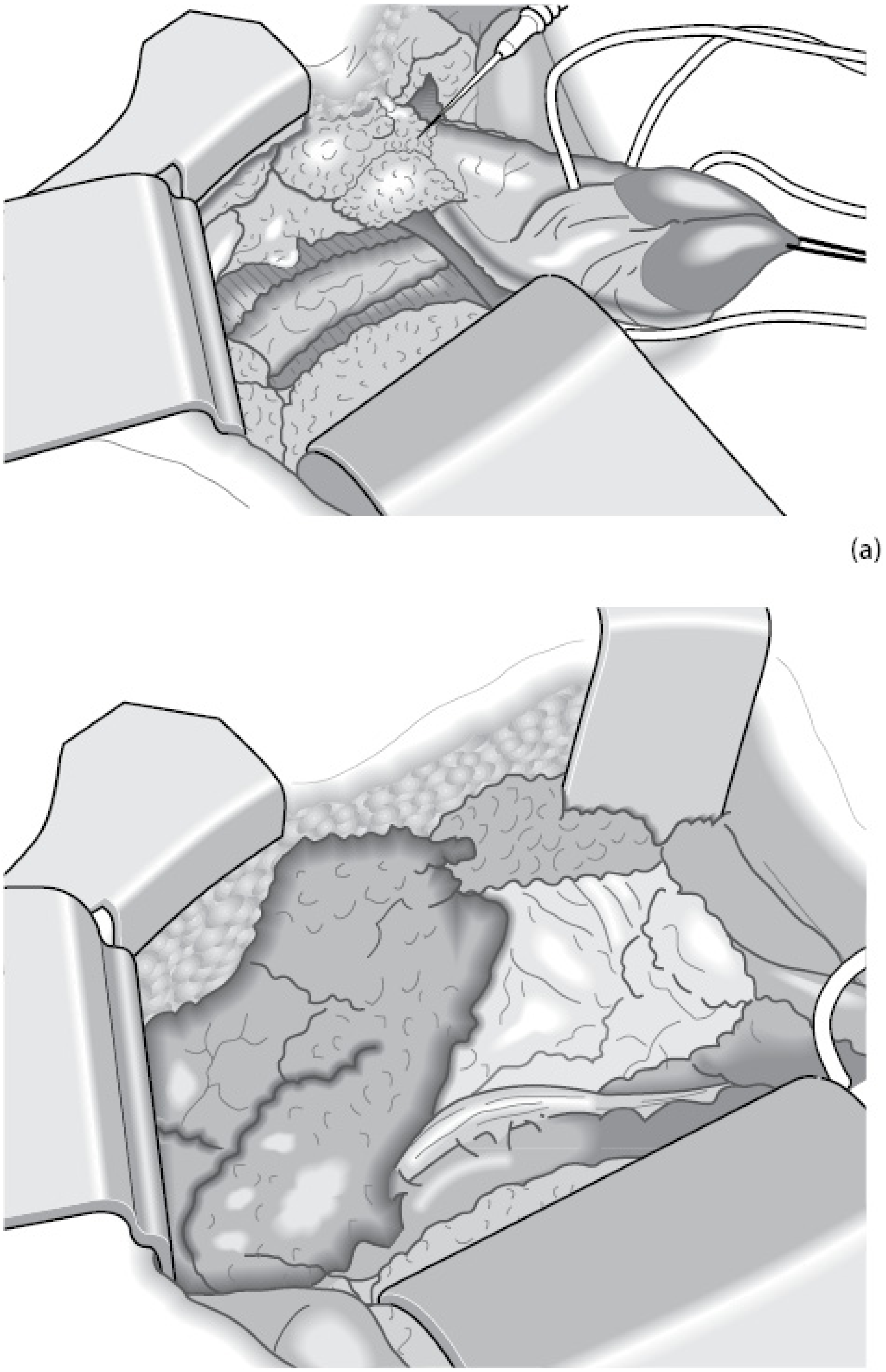
To mobilize the corpora, their neurovascular supply (the pudendal artery, veins, and nerve) must be freed along their course from behind the spine of the ischium from where they run forward in Alcock's canal. This is a fascial channel that runs forward on the surface of the internal obturator fascia, lateral to the ischiorectal fossa, below the pelvic floor (**Figure 79.19a**). A retroperitoneal plane lateral to the bladder is developed by forefinger dissection behind the lower rectus toward the pelvic floor. Medial retraction of the bladder and rectum behind it reveals the pelvic floor muscles and a muscle stimulator helps to define their attachment to the obturator fascia (the so-called “white line”) (**Figure 79.19b**). The muscle fibers are divided about 0.5 cm medial to the white line with bipolar diathermy. This exposes the ischiorectal fat, which can be pushed downward and medially to expose the fascia over the internal obturator muscle (**Figure 79.19c**).


Pelvic floor muscle
Rectum
Obturator internus muscle
Alcock's canal
Ischiorectal fossa fat
(a)
(b)
(c)

79.19a–c

RELEASING THE BASE OF THE PENIS AND EXPOSING THE PEDICLE

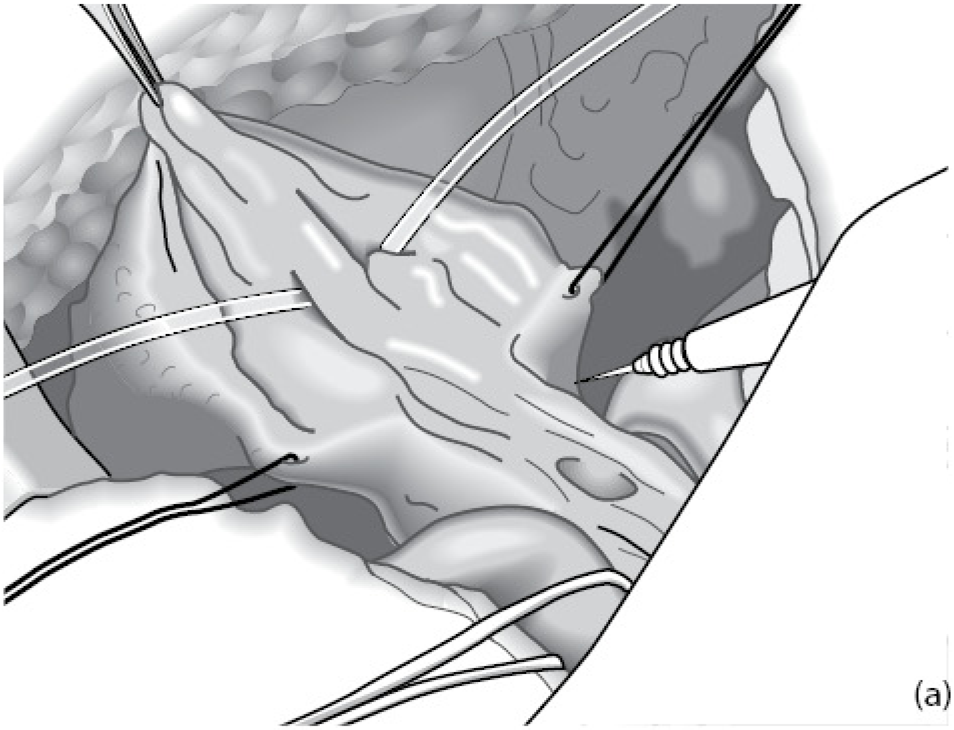
This dissection is continued anteriorly toward the pubic ramus, where the muscle is often thicker. The corporal body is now released from the bone by first incising along the inferior pubic ramus, lateral to its attachment (**Figure 79.20a**). A combination of blunt and bipolar dissection peels the periosteal attachment of the corpus off the bone. By careful dissection from above and below, the corpus is detached, and the pedicle can be demonstrated running from behind and along it (**Figure 79.20b**). Once it is seen, any fascial attachments lateral to the pedicle are divided, exposing fat and releasing the neurovascular bundle and the corpus medially.



79.20a,b

BLADDER NECK, BLADDER, AND PROXIMAL URETHRAL CLOSURE

The verumontanum is identified proximally on the urethral plate. Proximally, the area of the bladder neck is identified by marking and excising two triangles of bladder mucosa on either side (**Figure 79.21a**). The bladder neck is reconstructed around a 10 Fr silicone stent whose proximal end is secured to the bladder mucosa with a fine, absorbable suture (**Figure 79.21b**). Two layers of interrupted 4/0 Monocryl™ sutures are used and these are continued proximally in a single layer to close the bladder. A 12 Fr Malecot catheter is inserted into the bladder to be used for suprapubic drainage and the two stents are brought out through the midline closure (**Figure 79.21c**). The closure is continued anteriorly along the urethral plate.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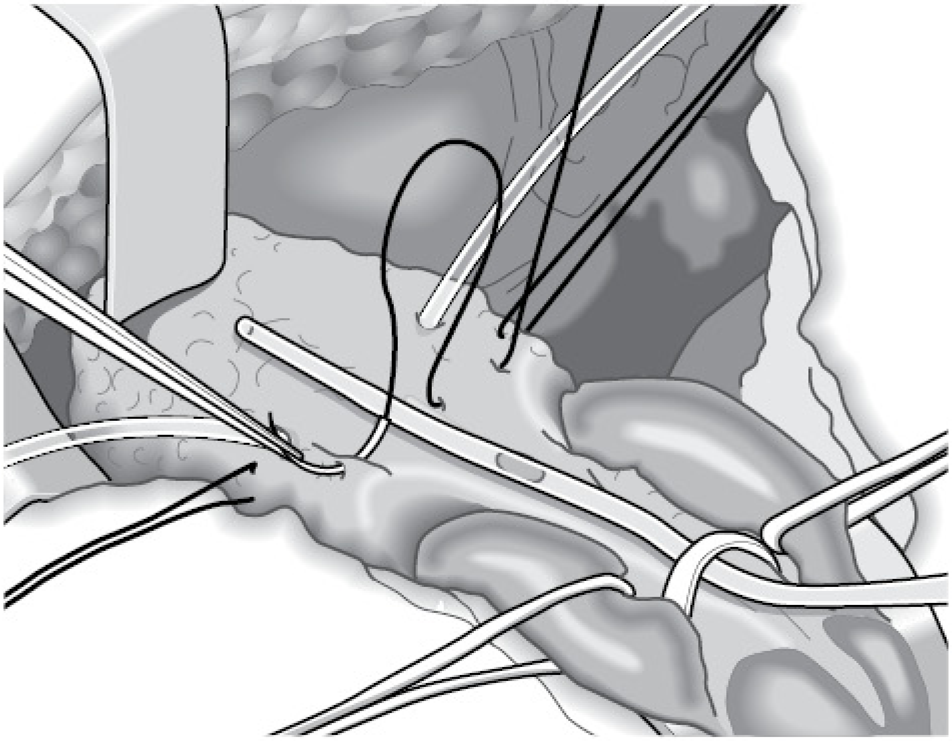
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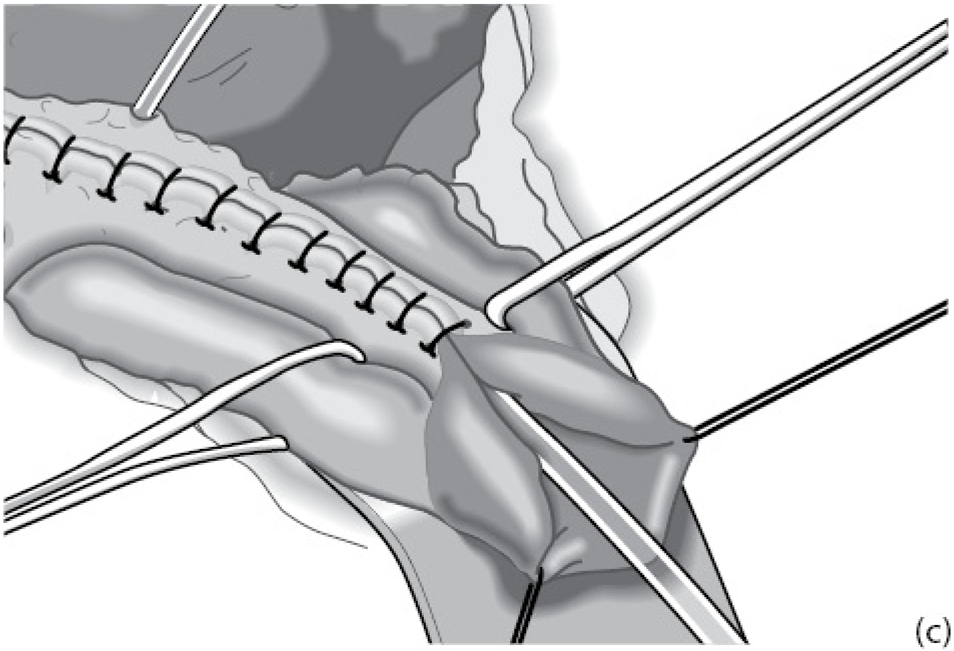


79.21a–c

CLOSURE OF URETHRA AND EPISPADIAS REPAIR

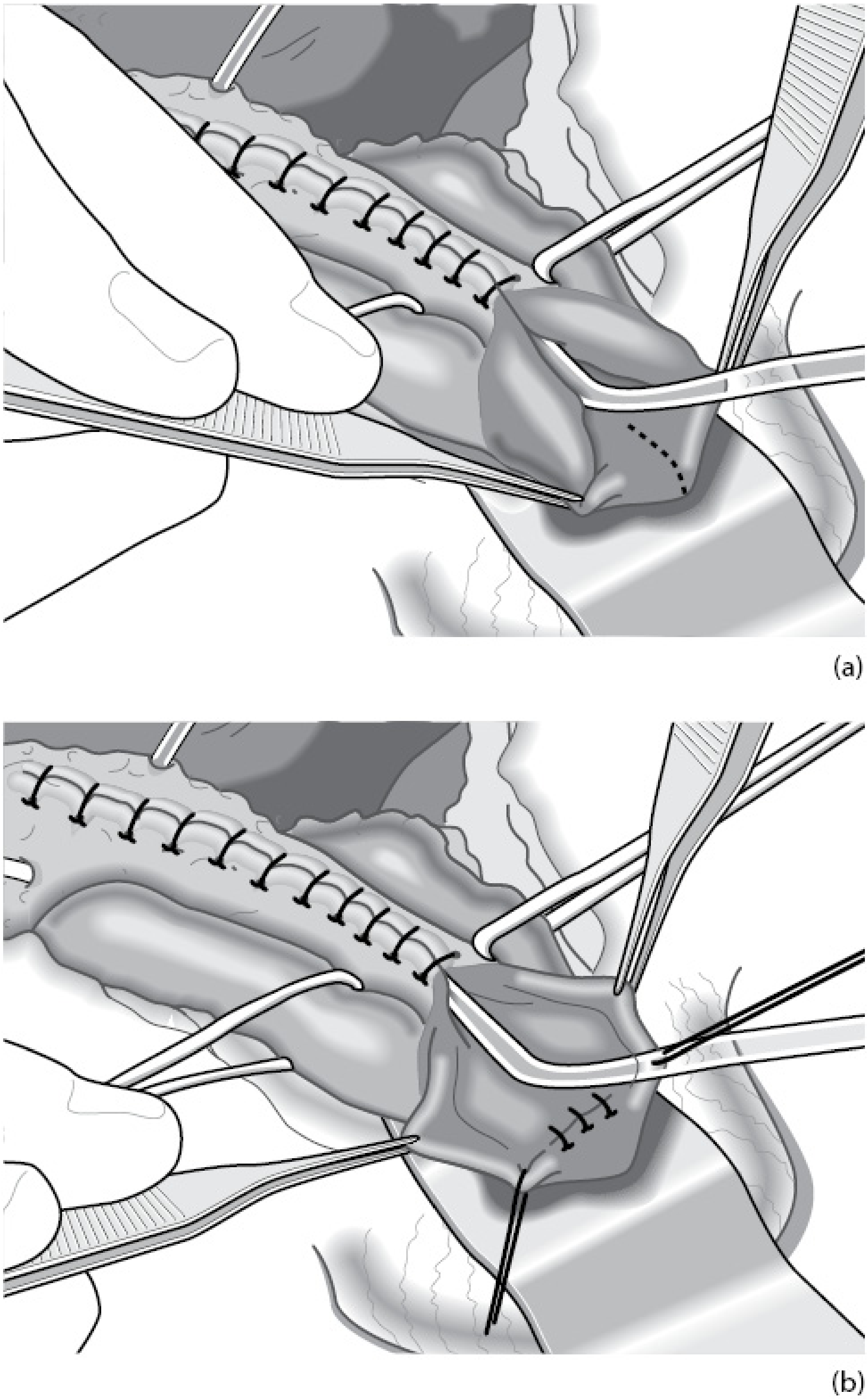
Option 1: Cantwell–Ransley repair

If the penis is a good length and protrudes well, a one-stage repair of epispadias can be undertaken.

"IPGAM" MANEUVER

IPGAM is the reverse of the meatal advancement and glanuloplasty (MAGPI) maneuver!

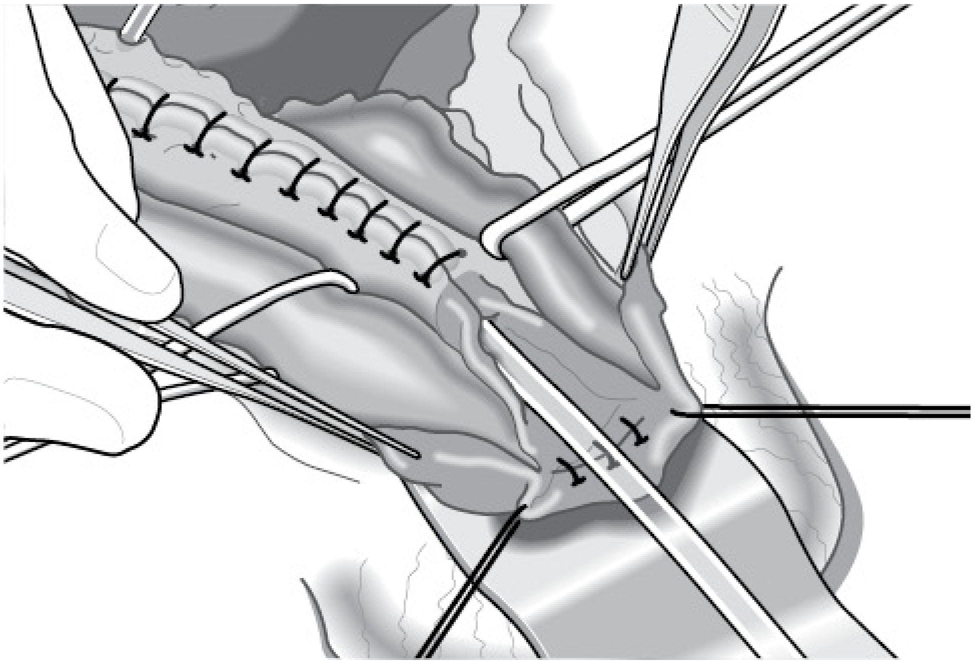
The distal urethral plate at the tip of the glans is incised longitudinally and the incision is closed transversely with fine, absorbable sutures (**Figure 79.22a and b**). This "IPGAM" maneuver brings the urethral meatus more ventrally on the glans.



79.22a,b

CLOSURE OF THE URETHRA AND GLANS

Two triangles of mucosa are marked and excised from either side of the glans, preserving the central strip of urethral plate (**Figure 79.23a**). Proximally, the plate is lifted from the corpora to provide greater mobility, which will allow the urethra's displacement ventral to the corpora and closure of the glans behind it (**Figure 79.23b**). The plate is then closed with interrupted 5/0 PDS™ sutures. The glans is reconstructed dorsal to it with two layers of interrupted, subcuticular 5/0 PDS™ sutures (**Figure 79.23c**).



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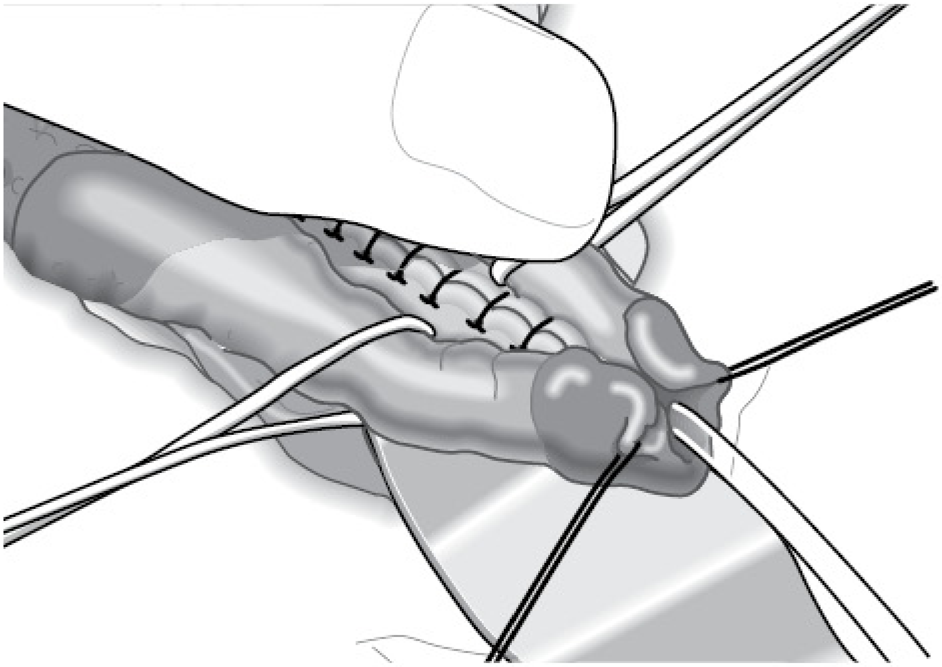

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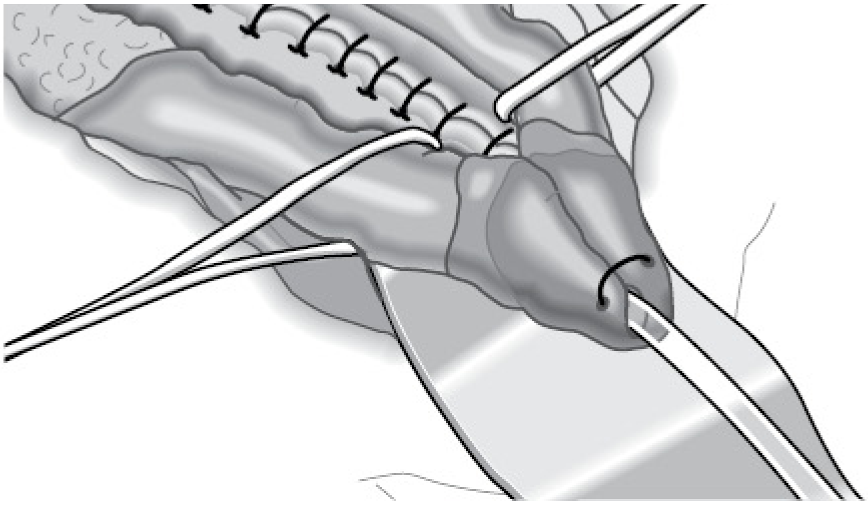
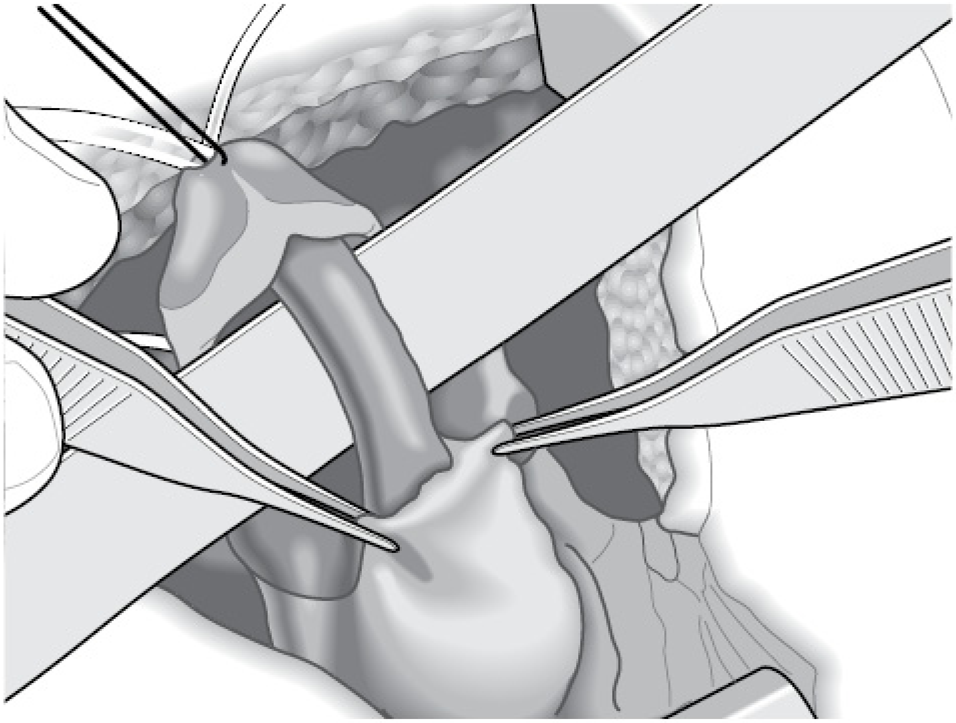

(c)

79.23a–c

CREATION OF THE SPHINCTER

The urethra is now free to bring ventral to the corpora, and this can be demonstrated by inserting a copper retractor (**Figure 79.24a**). The muscle between the corpora, at and below the level of the prostate, is again identified with the muscle stimulator. A sling of this muscle is wrapped around the urethra by passing two 4/0 Monocryl™ sutures on either side from behind. The wrap is not circumferential and the sutures should be tied dorsally, so as to be snug but not tight. The two corporal bodies are now sutured together in the midline with 4/0 Monocryl™ (**Figure 79.24b**). External rotation of each at this time will help correct the dorsal chordee.



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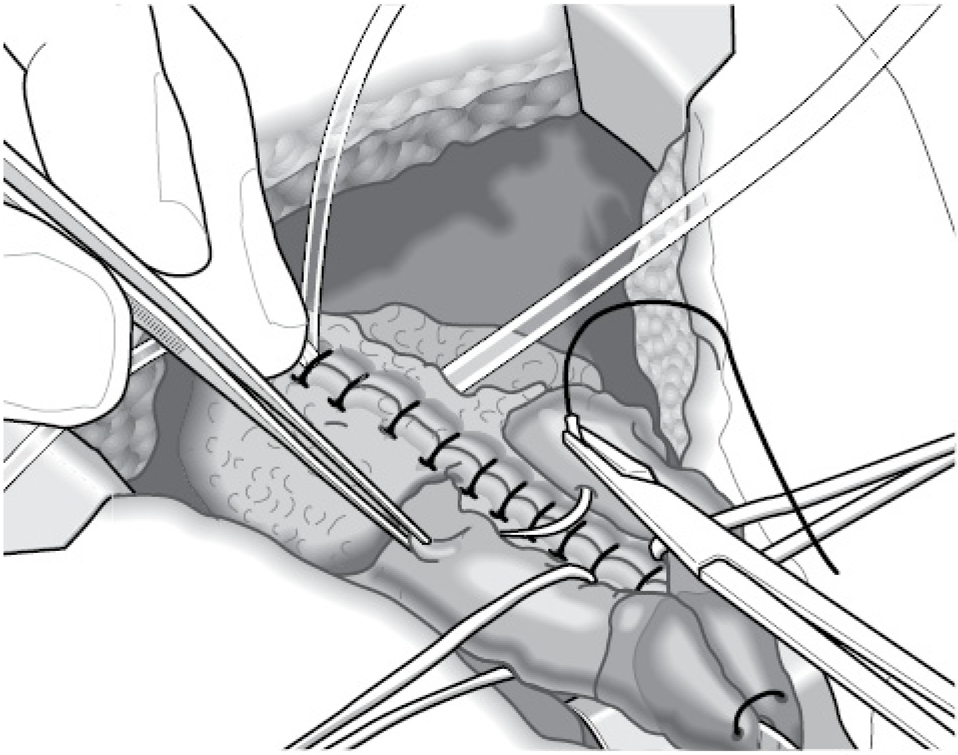
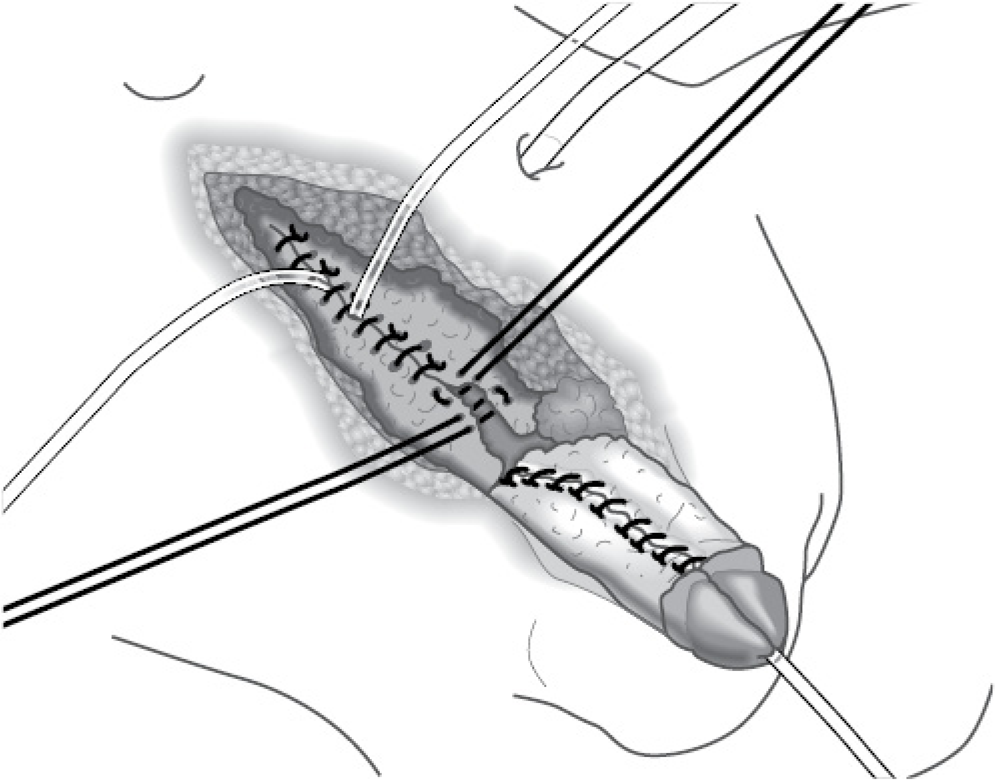


(b)

79.24a,b

CLOSURE

The abdominal wall is closed from above with figure-of-eight sutures of 2/0 PDS™, as for primary closure. The stents are brought out through the wound, while the Malecot is tunneled out to the left of the midline. Two 0 or 1/0 PDS™ sutures are placed at the lower end of the wound to reclose the symphyseal area. The base of each corpus (corresponding to the area previously separated from the bone) is sutured to the lower end of the abdominal closure with 3/0 PDS™ sutures (**Figure 79.25**). Care must be taken to avoid damaging the pedicle at this point.



79.25

PENILE SKIN COVER AND SKIN CLOSURE

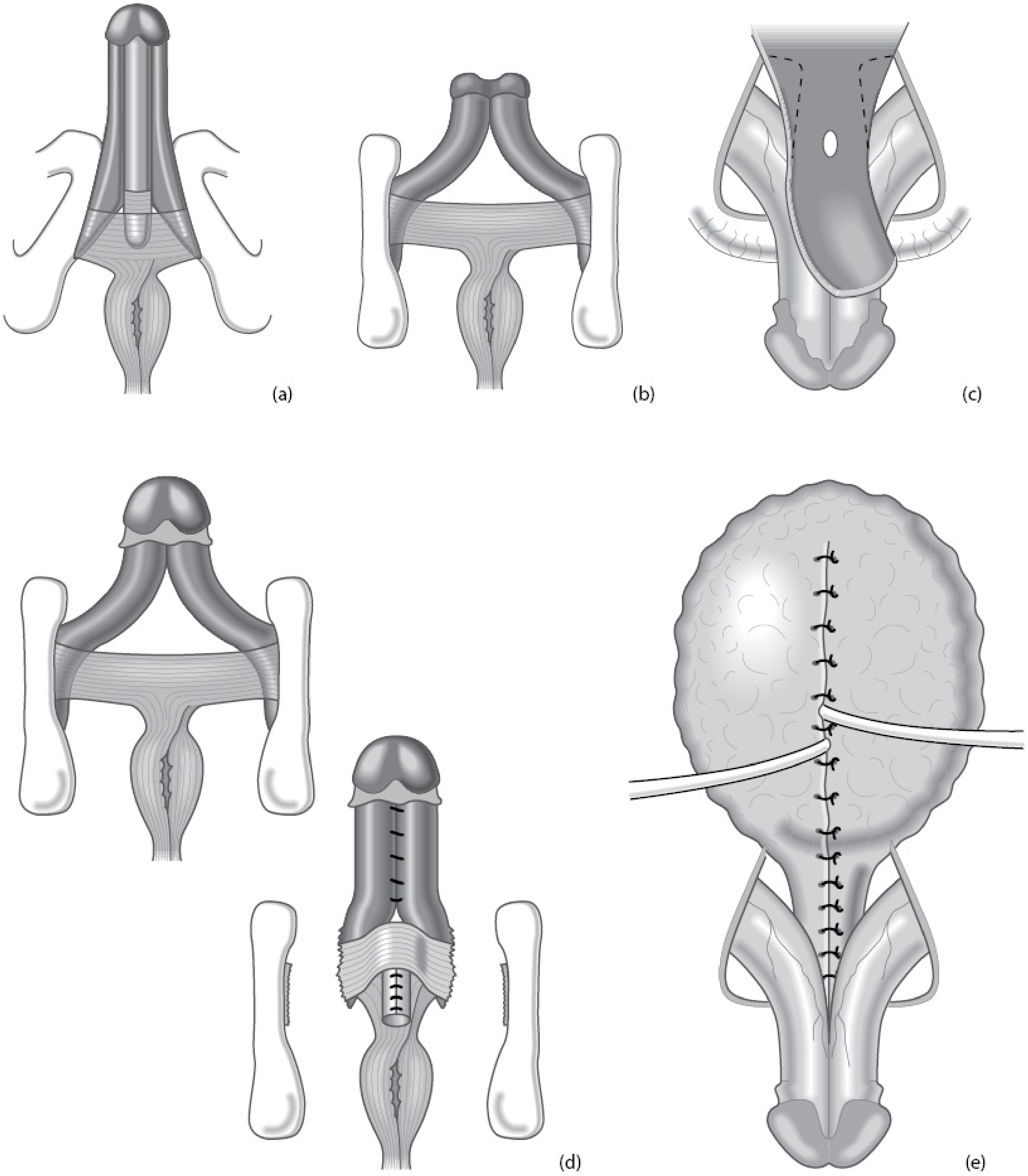
The penile skin flap is now lifted and flattened. It is incised in the midline and the apex of this incision is sutured to the frenular area of the skin cuff with 6/0 Monocryl™. The penis is wrapped and the skin closed dorsally in the midline or with Z-plasties. The edges are trimmed before the circumcision wound is closed with 6/0 Monocryl™. The subcutaneous layers and skin of the abdominal wall are closed with absorbable suture.

Option 2: Creation of hypospadias

Where the penile length is compromised (as is often the case) and when the urethral plate appears to limit penile length, this approach is the better one to use. In practice, surgeons have more experience with this approach and the published data on continence are principally from these patients. Subsequent procedures are required to create the distal urethra, with or without skin grafting.

LIFTING OF THE URETHRAL PLATE FROM THE CORPORA AND GLANS

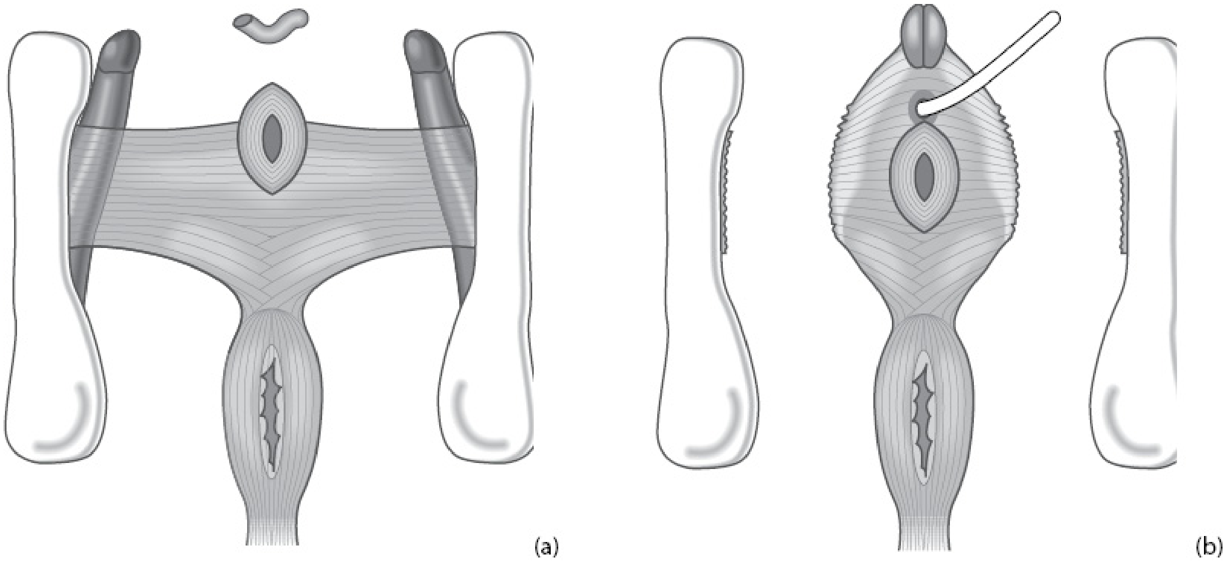
The urethral plate is separated completely from the corpora, including its glanular section. This is tubularized throughout its length with interrupted 5/0 Monocryl™. The neourethra is then dropped between the two corporal bodies. From the ventral aspect, the perineal muscle is identified and wrapped around the urethra using interrupted 4/0 Monocryl™. The wrap is not complete and the sutures are snug but not tight. The corpora are joined dorsally and the penile shaft reconstruction is completed as above, with corporal rotation to correct chordee. The two hemiglans are joined in layers with 5/0 PDS™ sutures. The urethral opening is secured to the ventral shaft with 5/0 PDS™ sutures. When the skin is reconstructed, a midline opening is made to accommodate the hypospadiac meatus, which is secured around its circumference with absorbable sutures (**Figure 79.26a–e**).



79.26a–e

FEMALE EXSTROPHY/EPISPADIAS REPAIR

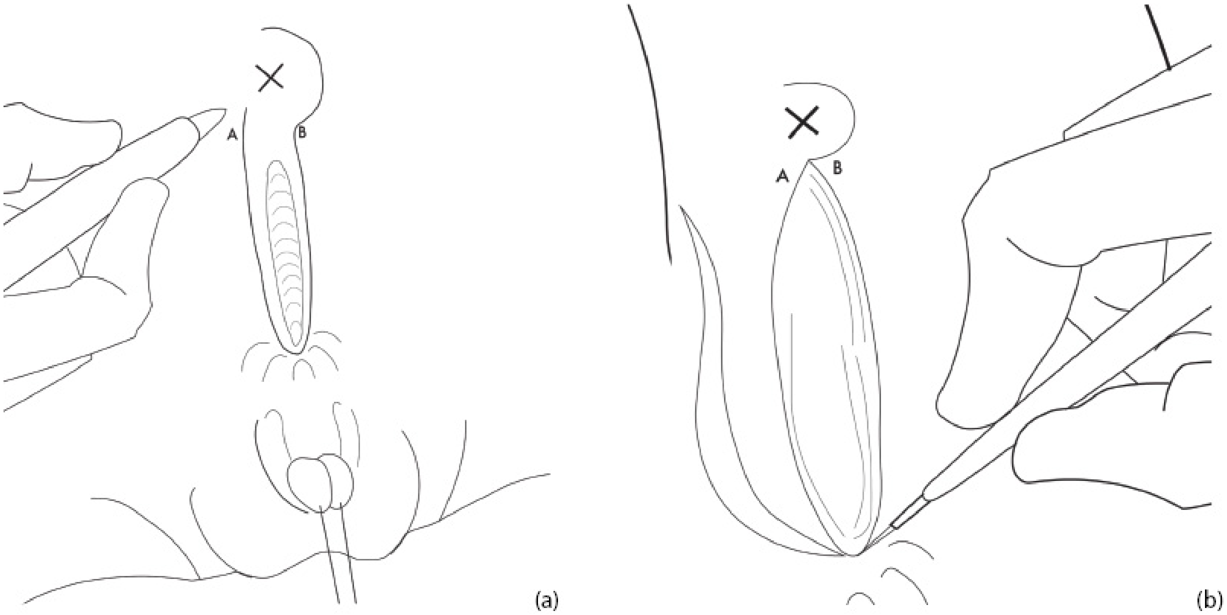
In females, after successful primary exstrophy closure or for primary epispadias, soft-tissue reconstruction is used to enable continence and to improve genital appearance. The clitoral corpora are dissected as for the penile corpora, with the labia minora skin flaps left attached around the glans for subsequent reconstruction (**Figure 79.27a**). The pelvic floor and pudendal pedicles are dissected in the same way and the muscle is identified below and lateral to the vagina—attached to the base of the corpora on either side. At the time of bladder outlet reconstruction, this muscle is brought around the dorsum of the proximal urethra and effectively wraps both the urethra and vagina (**Figure 79.27b**). At closure, the hemiclitori can be brought together and the labia minora replaced lateral to the neourethral orifice and the vagina. The mons is reconstructed by mobilizing subcutaneous fat from either side.



79.27a,b

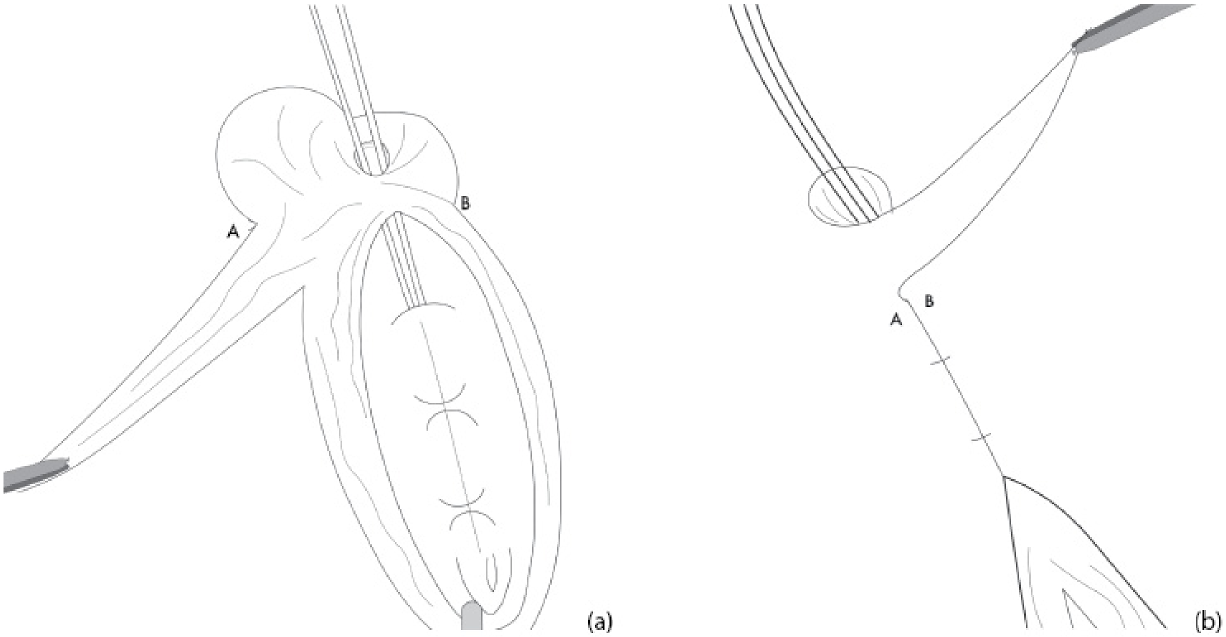
UMBILICOPLASTY

A cosmetic umbilicoplasty can be achieved by prior planning of skin incisions to create a spiral flap. The position of the neoumbilicus is marked at the start of the operation at the level of the iliac crests. The two proximal ends of the longitudinal incision, labeled, A and B in **Figure 79.28a**, will be approximated together at closure. A double incision is made to make a distal flap with greater width of skin on the right side of the wound (**Figure 79.28b**).



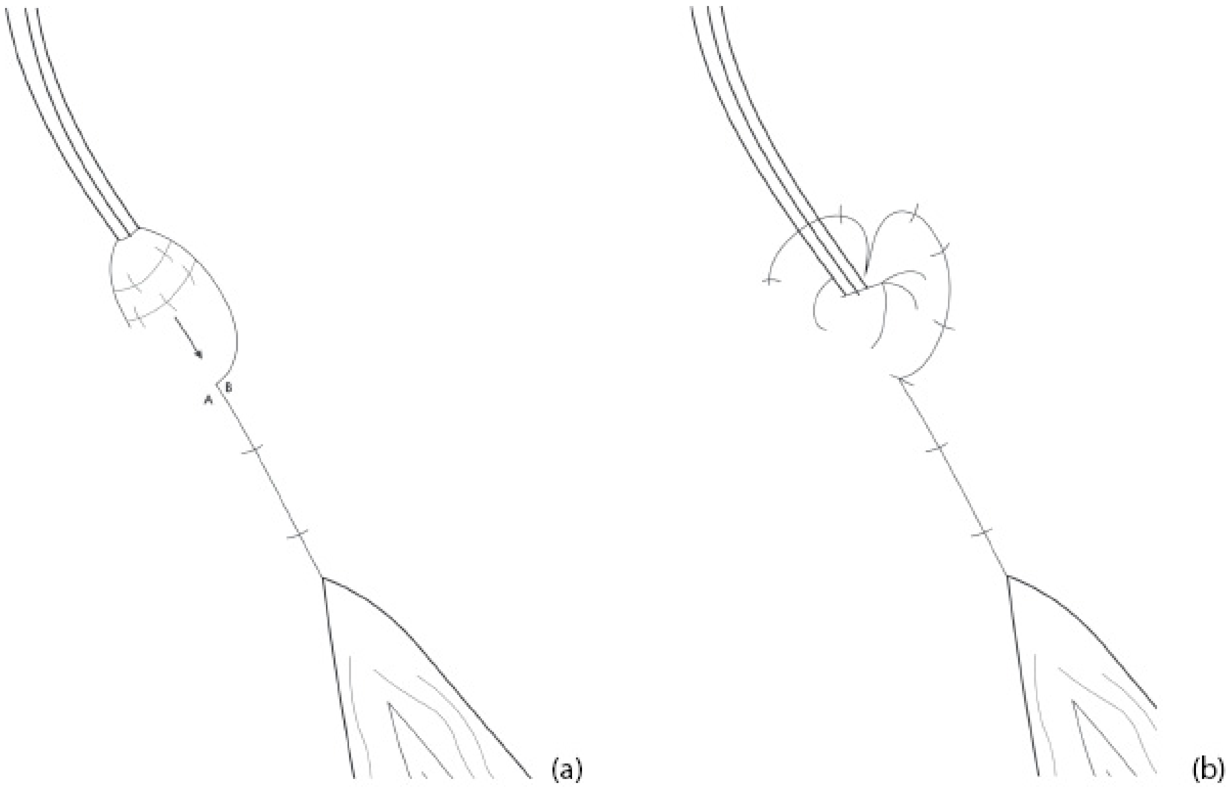
79.28a,b

At the umbilical level, a 240-degree disk of skin is lifted in continuity with the flap (**Figure 79.29a**). When the abdomen is closed at the end of the procedure, the flap is lifted and fat and scar tissue excised from beneath. A well is created in the fat at the umbilical level and the ureteric catheters brought through it. The flap is spiraled around the catheters in an anticlockwise fashion, joining the corners A + B below the umbilicus and bringing the wider strip of normal skin to form the left-hand edge of the umbilicus (**Figure 79.29b**).



79.29a,b

The completed cone is inverted around the catheters and its apex may be secured to the abdominal wall fascia before completing the wound closure (**Figure 79.30a and b**). This creates a neoumbilical appearance upon recovery from the Kelly repair.



79.30a,b

Postoperative care

After radical soft-tissue reconstruction and epispadias repair, the penis is enclosed in a foam or adhesive dressing for 1 week. The ureteric stents are left on drainage and removed after 1 week, and the bladder remains on free drainage for a further 3 weeks. At this time, the urethral stent is removed and the suprapubic Malecot catheter is clamped intermittently. When this is possible for 3 hours and the child has voided urethrally (checked by US), the suprapubic catheter is removed. Continence and bladder capacity will evolve with time, and regular evaluations at approximately 3-month intervals are required. Cystoscopic evaluation of the bladder outlet is performed with a fine cystoscope (8 Fr) at 3–6 months.

FURTHER READING

Featherstone NC, Cuckow PM. Spiral rotational flap for the creation of a new umbilicus in bladder exstrophy. *J Pediatr Urol* 2015; 11: 96–7.

Gearhart JP. The bladder exstrophy–epispadia–cloacal exstrophy complex. In: Gearhart JP, Rink RC, Moriquand P (eds). *Pediatric Urology*. Philadelphia, PA: Saunders; 2001: pp. 511–46.

Kelly JH. Exstrophy and epispadias: Kelly's method of repair. In: O'Neill JA, Rowe MI, Grosfeld JL, et al. (eds). *Pediatric Surgery*, 5th ed. St Louis, MO: Mosby; 1998: pp. 1732–59.

Mushtaq I, Garriboli M, Smeulders N, et al. Primary bladder exstrophy closure in neonates: Challenging the traditions. *J Urol* 2014; 191: 193–7.

Siffel C, Correa A, Amar E, et al. Bladder exstrophy: An epidemiologic study from the International Clearinghouse for Birth Defects Surveillance and Research, and an overview of the literature. *Am J Med Genet C Semin Med Gent* 2011; 157(4): 321–32.

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Surgical treatment of disorders of sexual development

RAFAEL V. PIERETTI AND PATRICIA K. DONAHOE

INTRODUCTION AND DEFINITIONS

A child born with disorders of sexual development (DSD) must be evaluated immediately at birth by a well-organized and experienced medical team including pediatricians, neonatologists, pediatric endocrinology, pediatric surgeons/urologists, geneticists, and psychologists aiming to make, as early as possible, an accurate diagnosis and treatment plan of those syndromes than can affect later sexual development. However, only four result in DSD at birth. These include 46,XX DSD (formerly known as female pseudohermaphroditism, congenital adrenal hyperplasia, or adrenogenital syndrome), ovotesticular DSD (true hermaphroditism), 46,XY DSD (previously known as male pseudohermaphroditism resulting from undervirilization of an XY male), mixed gonadal dysgenesis (MGD) 45,X/46,XY, or the rare 46,XY patient with pure gonadal dysgenesis having a 46,XY karyotype but a female phenotype. The gender, which is appropriate to the anatomy of the infant, must be decided as early as possible without delay, since new parents are asked about the sex of the child immediately after birth. They must be able to give an answer that is commensurate with the gender assignment that will eventually provide the most satisfying functional result. However, it is important that parents are reassured and to work closely with a team of

DSD experts who will help the family make a decision with which all will be most comfortable.

Two practical screening criteria, gonadal symmetry or asymmetry and the presence of a Y chromosome, can be used to help in the early diagnosis of the infant as having one of the four DSD disorders. Probes for SRY can be used to sequence or genotype the *SRY* gene. Gonadal symmetry is determined by the position of one gonad relative to the other, either above or below the external inguinal ring. If both gonads are symmetrical, then a diffuse biochemical cause underlies the abnormality, as in 46,XX DSD female with excess androgens or in a 46,XY DSD male with diminished androgens, which are biochemical anomalies that influence both gonads equally. Asymmetry occurs in MGD and ovotesticular DSD (true hermaphroditism), which are chromosomal abnormalities in which a predominant testis descends and a predominant ovary remains above the external ring. The karyotype is always 46,XX in cases of 46,XX DSD (female pseudohermaphroditism with adrenogenital syndrome) and is most often (90%) the case in patients with ovotesticular DSD (true hermaphroditism). Patients with 46,XY DSD (male pseudohermaphroditism) or MGD always have a Y chromosome in their karyotype (see studies from MacLaughlin and Donahoe in 2004 and Donahoe and colleagues in 2005 in “Further Reading”).

Following this initial evaluation, rapid analysis for elevated 17 hydroxyprogesterone to detect CYP21 disorders must be carried out in masculinized females with 46,XX with symmetrical undescended gonads. A detailed history and physical examination can be coupled with polymerase chain reaction (PCR), fluorescent *in situ* hybridization (FISH), or direct sequencing to detect mutations or deletions known to cause defects in the four major categories, and to define these disorders more fully and guide sex assignment.

Genetic females, no matter how severely virilized, should be raised as females. In genetic males, the gender assignment must be based on the infant’s anatomy, i.e. the size of the phallus, and not on the 46,XY karyotype. If the phallus is inadequate, one should strongly consider assignment to the female gender. In examining the phallus, one should consider the gestational age of the patient. The average penile length at 30 weeks' gestation is 2.5 ± 0.4 cm, increasing to 3.5 ± 0.4 cm at term, with a width of 1–1.5 cm. A term size below 2 ± 0.9 cm should cause concern. Failure to respond to a trial of exogenous testosterone is an important criterion in some cases. Exceptions, however, must always be made if the patient presents late, and has become fully committed to the male role, or in the event of a diagnosis of 5-α-reductase deficiency.

Female pseudohermaphroditism occurs when a genetic female (46,XX) is exposed *in utero* to androgens, either exogenous or endogenous as in the congenital adrenal hyperplasia syndromes. The phenotype can vary from clitoral enlargement alone to complete labioscrotal fusion and formation of an entirely normal male penis with a closed urethra formed to its tip. Cortisol deficiency can lead to salt wasting, which can be life-threatening unless replacement is instituted early after birth. All masculinized females have normal child-bearing potential and should be raised as females.

Male pseudohermaphroditism occurs in 46,XY genetic males with deficient masculinization of the external genitalia due to insufficient testosterone production, conversion, or inadequate target organ response. Many patients with male pseudohermaphroditism have been raised as males. However, if the female gender is chosen, gonadectomy should be done at the time of perineal reconstruction. The patient with an absent or rudimentary vagina usually requires only a clitoroplasty and labioscrotal reduction. The labioscrotal folds should be partially reduced during the first procedure and dilatation or a substitute vaginoplasty planned for the late adolescent or early adult years. Patients with testicular feminization in whom an introitus is often present may have this dilated with bougies at a later age to form a functional vagina.

True hermaphrodites have well-developed, non-dysgenetic male and female gonadal tissue in many combinations, i.e. a testis on one side and an ovary on the other, two ovotestes, or a normal gonad on one side and an ovotestis on the other. Although over 80% of these patients have a 46,XX karyotype, testicular tissue is present. The patient with a small phallus should be raised as a female. However, the final decision should be made following adequate discussion with the parents or guardians about various options and choices of how their child can be reared. The patient with a large phallus already committed as a male should be raised as a male. Gonads should be bivalved and biopsied longitudinally. The gonadal tissue commensurate with the sex of rearing (ovary tissue is peripheral, testicular is central) should be salvaged. Perineal Müllerian reconstruction should be accompanied by removal of Wolffian structures if the female sex is chosen. If the phallus is adequate for male gender assignment, ovarian and Müllerian structures should be removed, followed by hypospadias repair. Testicular prostheses can be inserted later, should the testicular tissue be inadequate.

MGD patients have dysgenetic gonads, retained Müllerian structures, internal and external asymmetry, and a mosaic karyotype, often 45,X/46,XY. The dysgenetic gonads can develop neoplasms such as gonadoblastoma or seminoma–dysgerminoma; therefore, in cases of asymmetric gonadal phenotype, the intra-abdominal or streak gonad should be removed. In general, the assignment of gender in children with MGD is the most difficult, and controversial, and deserves careful consultation. In some cases, consideration should be given to raise the child as a female with removal of the gonads, perineal reconstruction with flap vaginoplasty, with estrogen and progesterone replacement at adolescence. Alternatively, the patient may undergo complex repair of their severe hypospadias and be reared as a male. In either case, long-term follow-up will be needed, as gender reassignment has occurred regardless of initial gender designation.

If the patient is already committed to the male role, then hypospadias repair will be required. The gonads must be carefully observed for tumor development, which may occur as early as the newborn period.

SURGICAL TREATMENT OF UROGENITAL SINUS ANOMALIES AND DISORDERS OF SEX DEVELOPMENT

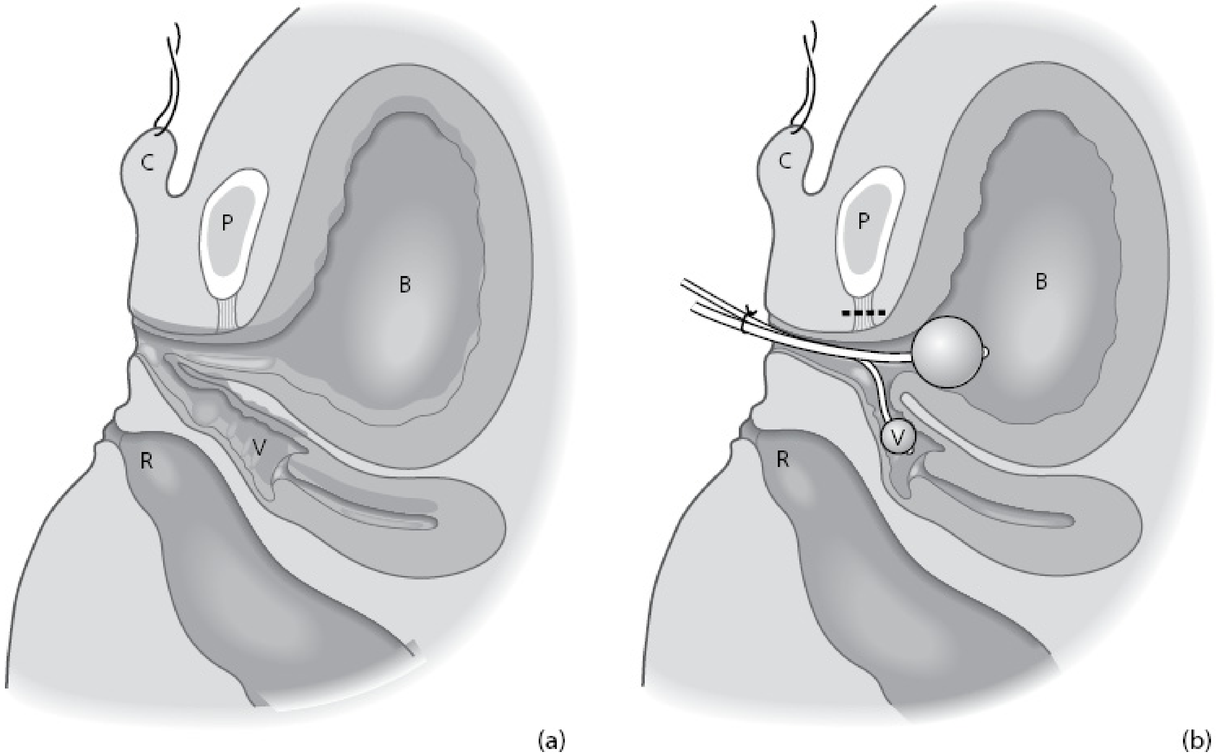
Patients with DSD and urogenital sinus (UGS) abnormalities are a source of great emotional stress for parents and patients, and present challenges for the surgical and medical or pediatric endocrinology teams involved in the patients' treatment.

There has been significant recent progress resulting from studies on clitoral innervation, most importantly the introduction of new nerve-sparing clitoroplasty techniques and the more widespread use of UGS mobilization procedures. However, there is a lack of sufficient long-term results regarding sexual function and acceptance of genital appearance in most patients, because most of the published studies have analyzed patients who have been reconstructed by operative techniques, which have been more recently modified.

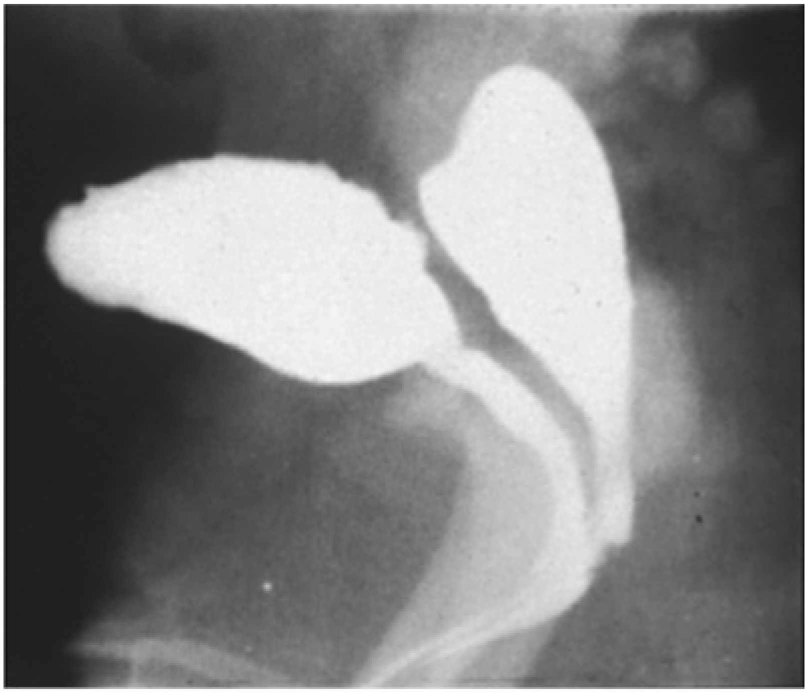
PREOPERATIVE EVALUATION

Imaging

A retrograde genitography is performed by occluding the opening of the UGS with the inflated balloon of a size 8 Fr Foley catheter placed outside the meatus and secured in place with tape (**Figure 80.1a and b**); lateral and oblique images are required. The catheter should be then advanced into the bladder for a voiding cystourethogram (VCUG) (**Figure 80.2**). In approximately 80% of the cases, the level of confluence of the UGS in relation to the bladder neck and external sphincter can be identified, thus facilitating the planning of the surgical procedure.

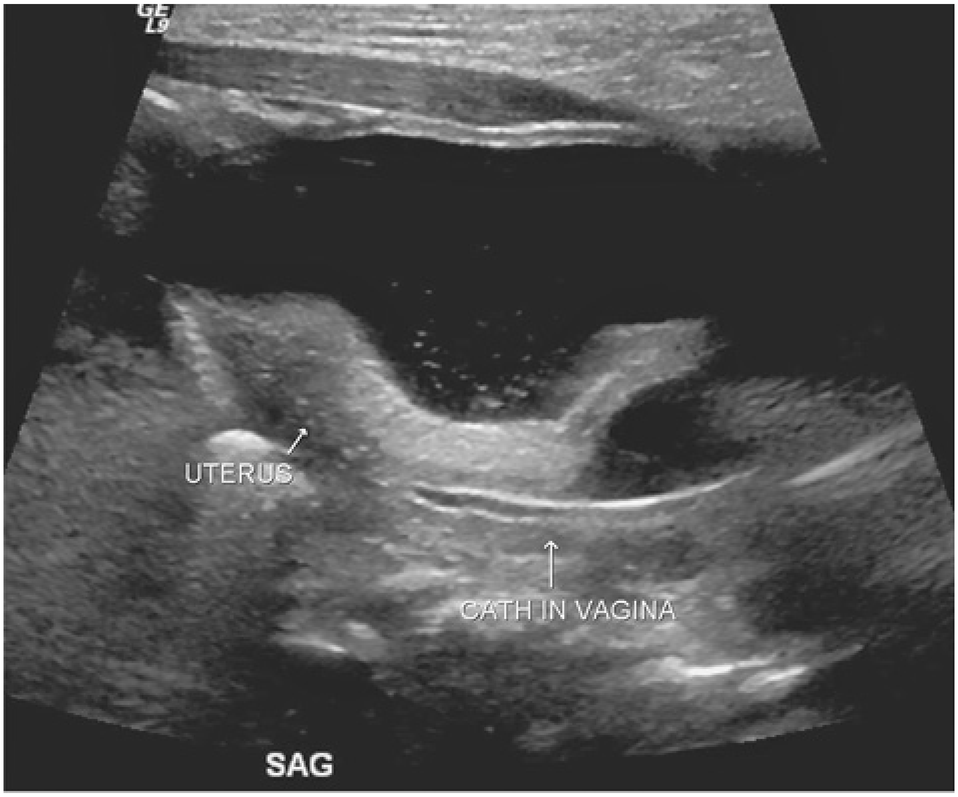

C
P
B
V
R
(a)
C
P
B
V
R
(b)

80.1a,b (a) Low confluence; (b) high confluence. (C, clitoris; P, pubis; B, bladder; V, vagina; R, rectum.)

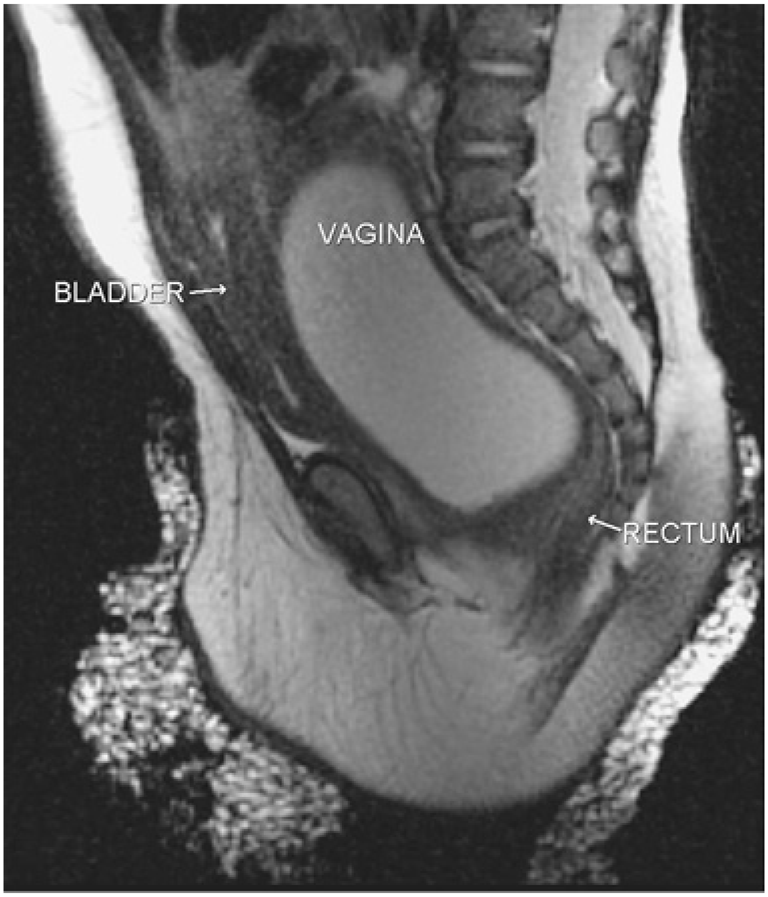


80.2 Voiding cystourethrogram showing low confluence.

Ultrasonography gives important information about the urinary tract, and the uterus, vagina, and gonads can be visualized (**Figure 80.3**). In those cases in which the anatomy is not well demonstrated, magnetic resonance imaging (MRI) of the pelvis can more clearly define the anatomy of the pelvic organs (**Figure 80.4**).


GE
L9
UTERUS
CATH IN VAGINA
SAG

80.3



80.4

Endoscopic evaluation

Endoscopy is of great help to define the anatomy for surgical reconstruction and it should be performed before and during the surgical reconstruction to corroborate the radiographic findings such as the location of the UGS and its distance from the bladder neck, inspection of the vagina to identify the cervix, bladder, and urethra as well as the distance from the vagina to the bladder neck. This highly important information is necessary to plan the type of surgical procedure.

Laparoscopy

Laparoscopy allows excellent visualization of the pelvic organs; it can be performed simultaneously with a gonadal biopsy or gonadectomy, and can be very helpful for identification or removal of Müllerian structures. In addition, laparoscopy can be used to perform other procedures, such as a laparoscopic-assisted sigmoid vaginoplasty, and for the removal of enlarged and symptomatic Müllerian duct remnants.

RECONSTRUCTION COMMENSURATE WITH FEMALE GENDER ASSIGNMENT

All 46,XX DSD newborns should be assigned to the female gender, regardless of the extent of masculinization, and should undergo surgical reconstruction concordant with the female gender assignment. Similar repairs can be used for selected patients who are not severely masculinized because of 46,XY DSD, MGD, or ovotesticular DSD. The key steps in a feminizing genitoplasty are clitoroplasty, labioplasty, or labioscrotal reduction and vaginoplasty, or mobilization and exteriorization. Surgical procedures must preserve clitoral sensation and excretion and result in normal-looking external genitalia, with a well-lubricated vagina, which will allow satisfactory and painless sexual intercourse. Due to some concerns regarding the benefits of clitoroplasty, it should be undertaken only after extensive discussions with the family, because another option is for deferral until the patient is capable of making her own decision.

Preoperative preparation

The bowel must be prepared before repair. For low repairs, magnesium citrate should be given starting 2 days before repair. For high repairs, a polyethylene glycol isotonic solution (Golytely; Braintree Laboratories, Braintree, MA, USA) is administered by mouth beginning 3 days before surgery for 2 consecutive days, followed by magnesium citrate 1 day before surgery. If needed, Ondansetron may be indicated to prevent nausea, or Golytely can be administered through a small nasogastric tube. The use of Golytely should be discontinued at least 24 hours before surgery to avoid leakage during the procedure. Magnesium citrate, which shrinks the bowel, is given on the last day to prevent leakage. Oral administration of neomycin plus erythromycin can be prescribed to reduce bacterial concentration although recent evaluations of the microbiome questions that practice.

Hormonal management of the 46,XX patient with adrenogenital syndrome is coordinated with experienced pediatric endocrinologists. Stress doses of steroids are given at the time of anesthetic induction. Steroids are continued during surgery and for 2–3 days after surgery at double the usual oral dose, followed by a tapering of the dosage. Patients on dexamethasone are asked to omit this medication, but children on prednisone take the usual morning dose on the day of surgery.

Planning and timing of surgical reconstruction

The magnitude and timing of surgical reconstruction are the subjects of significant controversy. Some groups advocate delaying sex assignment to an age in which each patient can make his or her own decisions; however, as mentioned earlier, most of these studies have been based on analysis of the outcomes of older surgical procedures. The authors believe that newer techniques result in an improved cosmetic appearance, a reduced complication rate, and are more likely to preserve sensation and function. In addition, except for MGD and delayed diagnosis of congenital adrenal hyperplasia (46,XX DSD), the sex of assignment is not ambiguous.

All available treatment alternatives are discussed with the parents and recommend that the different steps of the surgical reconstruction should be incorporated as a single surgical procedure and be performed at an early age in order to take advantage of all available tissues, and to achieve the best possible functional and cosmetic results. Patients with a low-confluence UGS can be operated on once their metabolic management is well controlled; in most cases, the authors undertake an elective reconstruction between 3 and 6 months of age, but repair can be done in the newborn period if the social situation so warrants. Patients with a mid-level or high confluence can be electively repaired at 9–12 months of age.

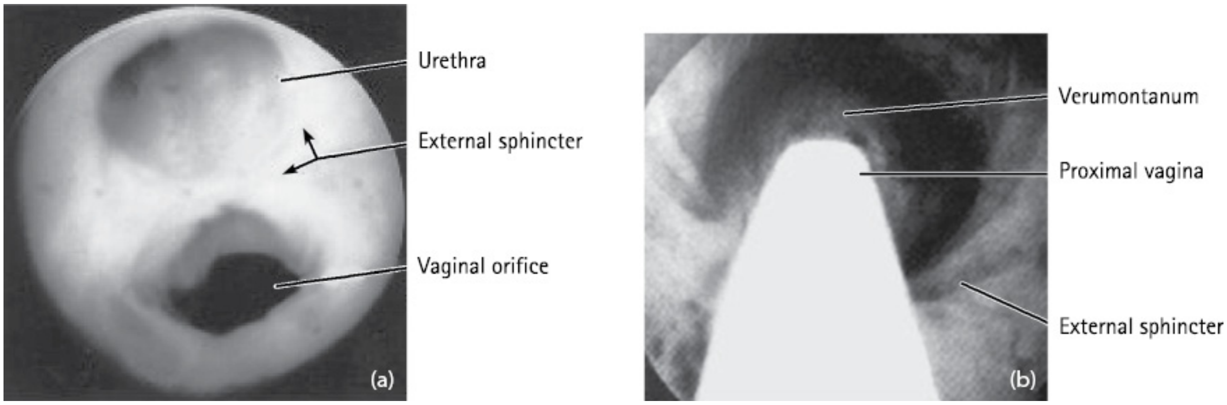
Adequate and controlled hormonal treatment is needed to prevent clitoral hypertrophy after correction.

Planning of the surgical reconstruction should incorporate the three components of a feminizing genitoplasty, in which the prepuce is used to create labia minora, if needed the clitoris is reduced, and the labiosacrotal swellings are used to fashion female-appearing labia majora and to enhance the vaginoplasty.

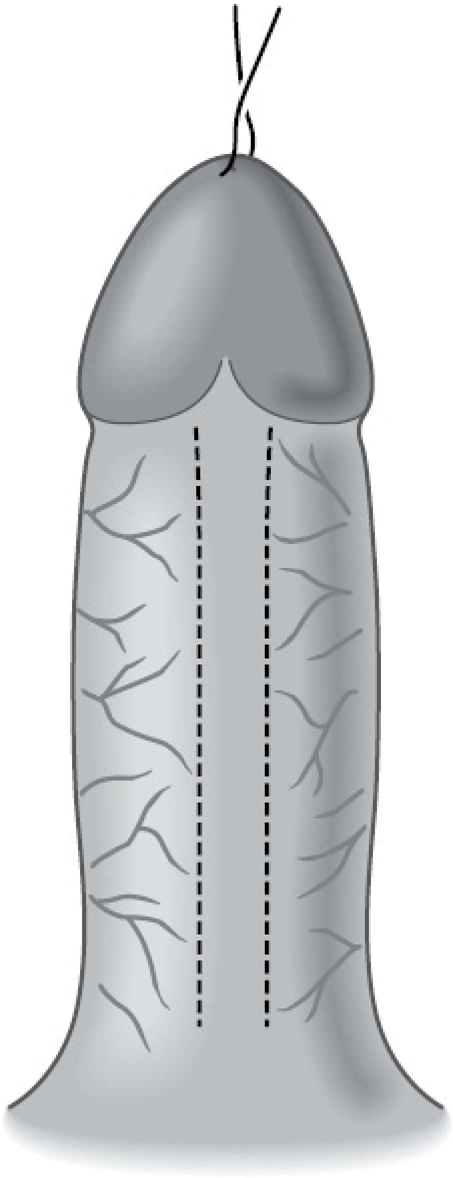
To improve surgical exposure, the authors use a hyperextended lithotomy position with the buttocks lying over and slightly beyond several folded towels.

Panendoscopy

All reconstructive procedures must begin with a panendoscopy using a pediatric cystoscope with 0-degree and 30-degree optics (**Figure 80.5**). High-flow irrigation of the UGS facilitates finding a very small vaginal orifice in the back wall of the UGS; in some cases, there may be only a pinpoint orifice, which can usually be found by probing with a 3 Fr ureteral catheter with a stiffened stent. For surgical planning, one must find the precise location of the confluence point between the vagina and urethra, and judge its relation with the bladder neck and the external sphincter. Those anomalies with the confluence point at or above the verumontanum/external sphincter are considered high, and those below are considered low. Patients with 46,XX DSD, MGD, and ovotesticular DSD, as well as those with 46,XY or 45,X/46,XY MGD have a cervix at the most proximal part of the vagina. Patients with 46,XY DSD have either a small prostatic utricle or a deeper, more generous cavity that has no proximal cervix. The prostatic utricle is characteristically found in the center of a flattened verumontanum, but has no surrounding prostatic tissue. In those patients with a mid-level and high confluence, a Fogarty catheter with a stopcock valve is passed into the vagina and the balloon is inflated; a small Foley catheter is also placed in the bladder, and both are labeled and tied together (see also **Figure 80.1a and b**).


Urethra
External sphincter
Vaginal orifice
(a)
Verumontanum
Proximal vagina
External sphincter
(b)

80.5a,b Endoscopic view of urogenital sinus.



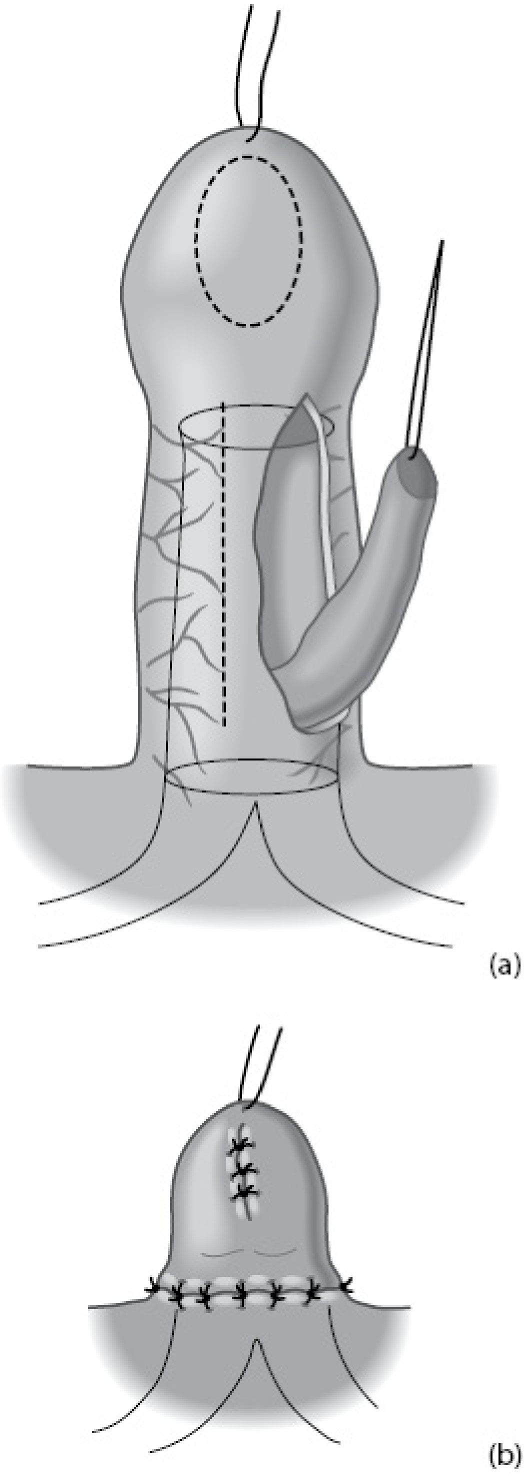
80.6

Clitoroplasty

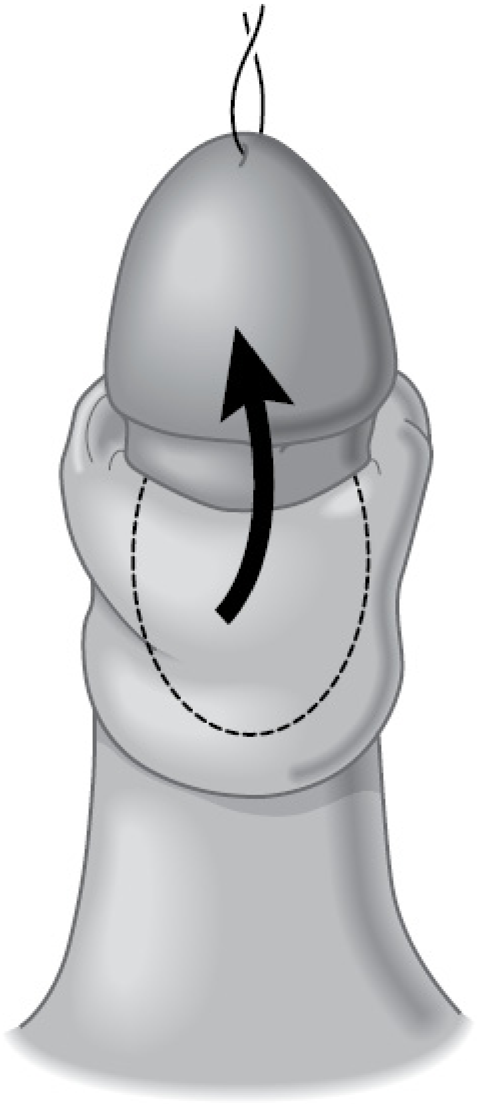
Clitoral resection and recession are of historical interest and no longer recommended. The goal of current techniques is to preserve sensation for future orgasms, provide an acceptable cosmesis, and avoid painful erections. A subtunical excision of the erectile tissue has been used extensively and led to newer nerve-sparing techniques. In 1999, Baskin et al. found that distribution of the sensory nerves of the clitoris, similar to the sensory nerves of the penis, is found on the dorsal aspect of the clitoris coursing under the pubis. Circumferential branches from the dorsal neurovascular bundle encircle the clitoral shaft toward the ventrum, thus making a ventral approach to the corpora most likely to avoid nerve injury. In Baskin's technique, corporal tissue proximal to the bifurcation is left intact. In cases with severe masculinization, the clitoris is too large, resembling a penis; in such cases, the authors discuss the anatomical characteristics with the parents and advise a clitoroplasty.

A 5/0 Prolene stay suture is placed in the glans. Two vertical incisions are outlined with a marking pen on each side of the urethral plate (**Figure 80.6**), and the meatus is circumscribed as for hypospadias surgery, taking care to leave a redundant segment of dorsal inner foreskin to fashion a hooded prepuce, thus preserving an important source of sensation. Prior infiltration of incisions with 0.25% Marcaine/1 : 200,000 epinephrine prevents excess blood loss. The vaginoplasty is incorporated into the surgical reconstruction outlining a wide base inverted "U"-shaped perineal flap based on the anus and using the ischial tuberosities as a landmark; the apex of this flap is advanced and placed at the estimated final location of the widened vagina. The clitoris is degloved; the ventral strip of the urethral plate should initially be kept intact. Following degloving of the dorsal skin, the distal portion of the UGS (urethral plate) is divided below the glans clitoris (**Figure 80.7a**). The urethral plate (UGS) is mobilized off the ventral aspect of the clitoral shaft downward to below the bifurcation of the corporal bodies. Next, clitoral reduction is carried out; the authors do not use a tourniquet as it has been observed that bleeding from the erectile tissues is not significant, particularly in infants, although it can be considerable in the older child. Longitudinal ventral incisions are made; the erectile tissue is dissected within the bodies. The body of the glans is sutured to the corporal body stumps with absorbable sutures (**Figure 80.7b**). The redundant dorsal tunica albuginea and neurovascular bundle are placed in a subcutaneous pocket above the pubic bone, taking care not to impair the neurovascular elements. The reduced clitoral shaft is covered with surrounding fat tissue, thus fashioning a normal-looking mons pubis.

In most cases, the authors try to avoid reduction of the glans clitoris, but in patients with severe enlargement, a wedge of glans tissue can be excised from the ventral aspect of the glans with careful reapproximation of the glans tissue with interrupted sutures of 6/0 PDS□. The dorsal mucosal collar should cover the glans partially, giving it a hooded appearance (**Figure 80.8**). The remainder of the dorsal prepuce is divided in the midline (Byars technique) and reattached to the mucosal collar as with hypospadias repair. The preputial skin wings are rotated inferiorly and incorporated lateral to the urethral plate to create labia minora.



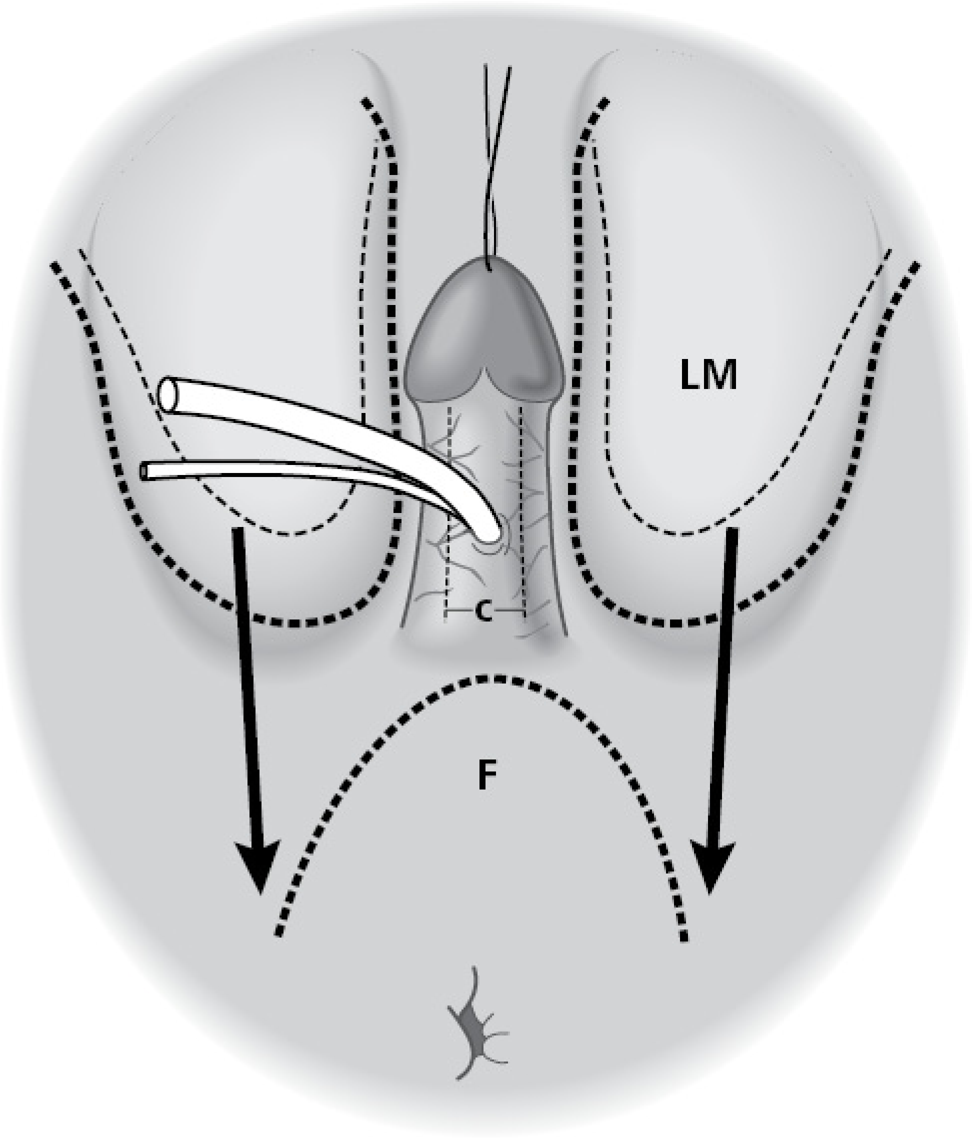
80.7a,b



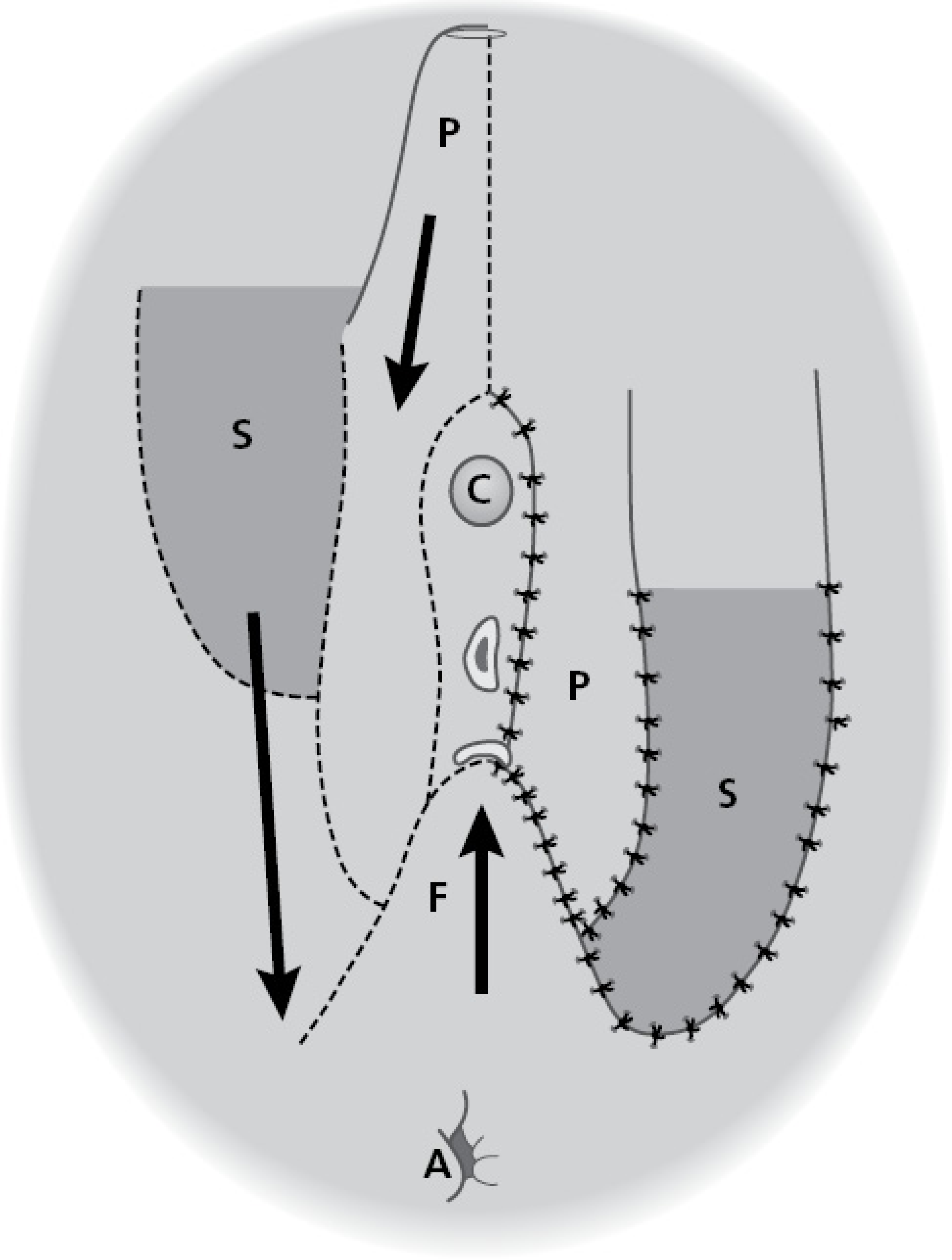
80.8

Labioplasty

Most girls with 46,XX DSD have labioscrotal swellings that are anterior in relation to the vagina than normal labia majora. Significant skin rugation may be present as well. To move this labioscrotal skin posteriorly, “Y”-shaped incisions are outlined with an extension posterior to the swellings. The scrotal flaps are cautiously defatted and moved posteriorly, beside the introitus, as bilateral Y–V advancements (**Figure 80.9**). The medial aspects of these skin flaps are then sutured to the lateral edges of the prepucial skin flaps mobilized during clitoroplasty (now labia minora) (**Figure 80.10**). The result is an anatomically correct positioning of the labia minora and majora posteriorly, beside the introitus, rather than anteriolaterally.



80.9



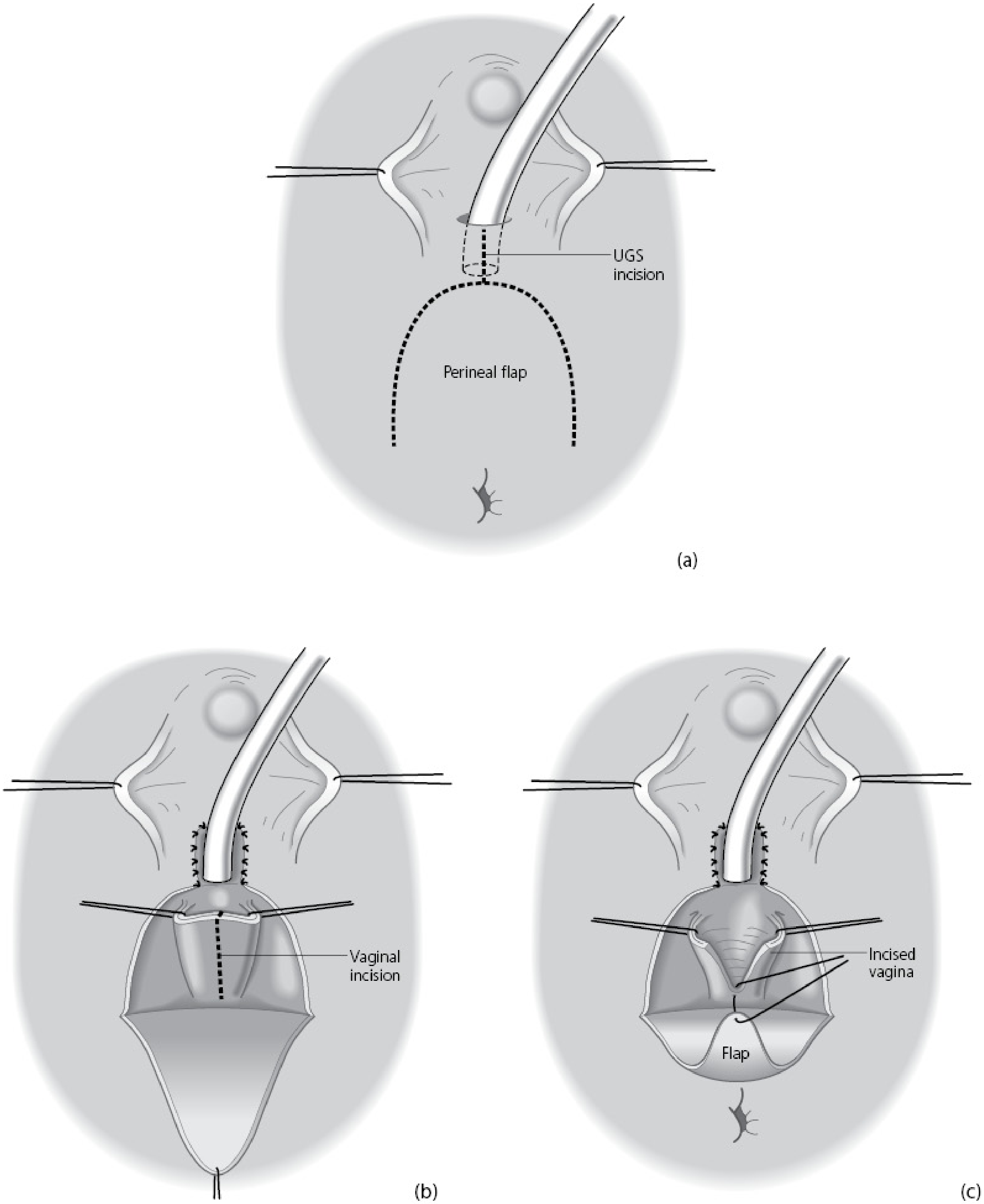
80.10

Vaginoplasty

The type of vaginoplasty is dictated by the anatomic location of the UGS confluence. There are six types of vaginoplasty: Cut-back, flap, pull-through, total and partial urogenital sinus mobilization, and vaginal replacement.

LOW-CONFLUENCE FLAP VAGINOPLASTY

Flap vaginoplasty should only be used in cases with a very low-confluence UGS, as otherwise it does not bring the merging point of the vagina with the urethra any closer to the perineum. The authors use a thick, inverted perineal skin "U" flap, based on the anus, which is outlined with a marking pen and which, when advanced, should reach the edge of the sinus (**Figure 80.11a**). The posterior wall of the vagina is dissected with care not to injure the rectum. Next, the posterior wall of the sinus must be opened longitudinally into normal caliber vagina to avoid a vaginal stricture (**Figure 80.11b**). The apex of the flap is inserted into the apex of the vaginal wall, and secured in place beginning with three interrupted, full-thickness sutures of 4/0 Vicryl™, which should be tied carefully to prevent tearing the fragile vaginal wall; the rest of the sutures are placed in a sequential manner (**Figure 80.11c**).

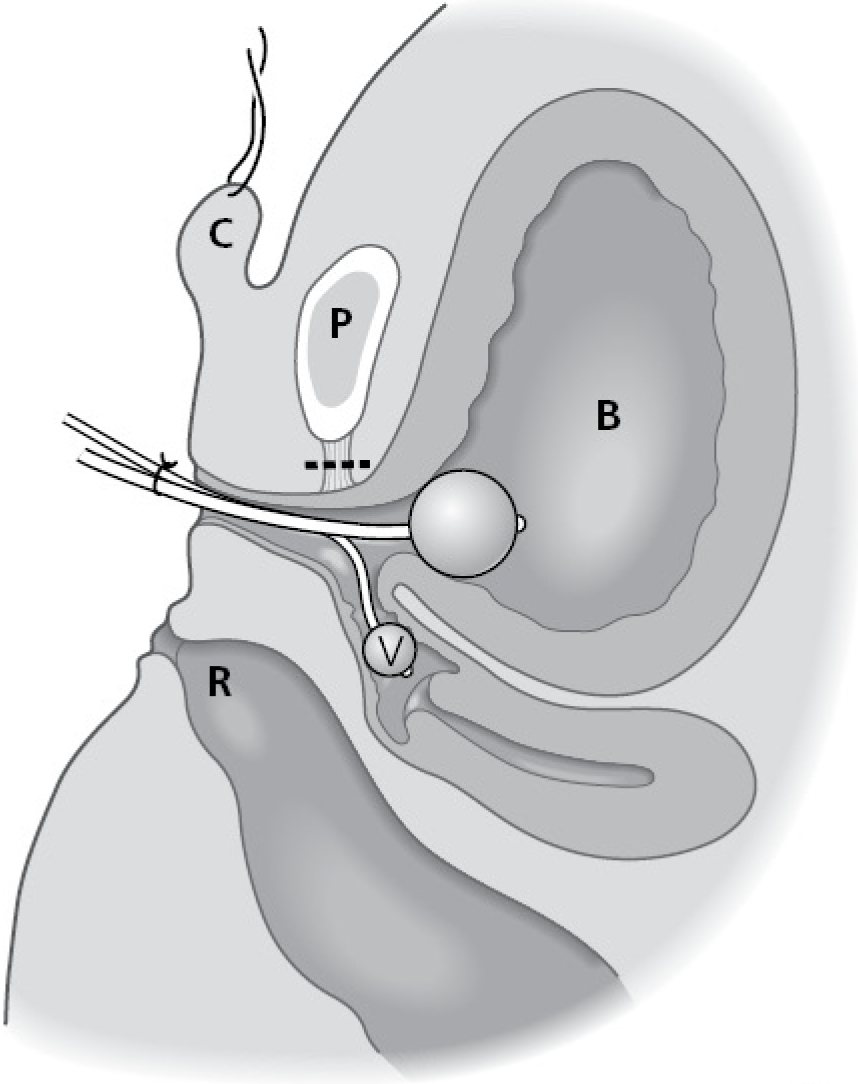

UGS incision
Perineal flap
(a)
Vaginal incision
(b)
Incised vagina
Flap
(c)

80.11a–c

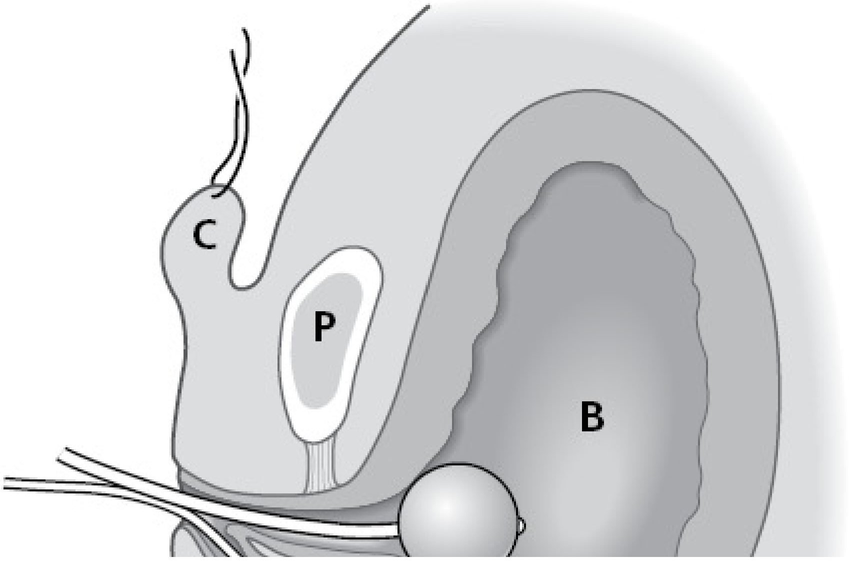
In low-confluence vaginoplasty, there is no need to insert a finger in the rectum, but a roll of Vaseline gauze can be inserted instead to avoid rectal injury. In low vaginoplasty, there is also no need to mobilize the UGS. Once the UGS has been dissected off the corpora, its ventral wall is opened longitudinally down to the vaginal opening to create a wet introitus. Later, during labioplasty, the medial edge of the prepuce (labia minora) will be sutured to the outer edge of the opened, non-mobilized urethral plate/UGS.

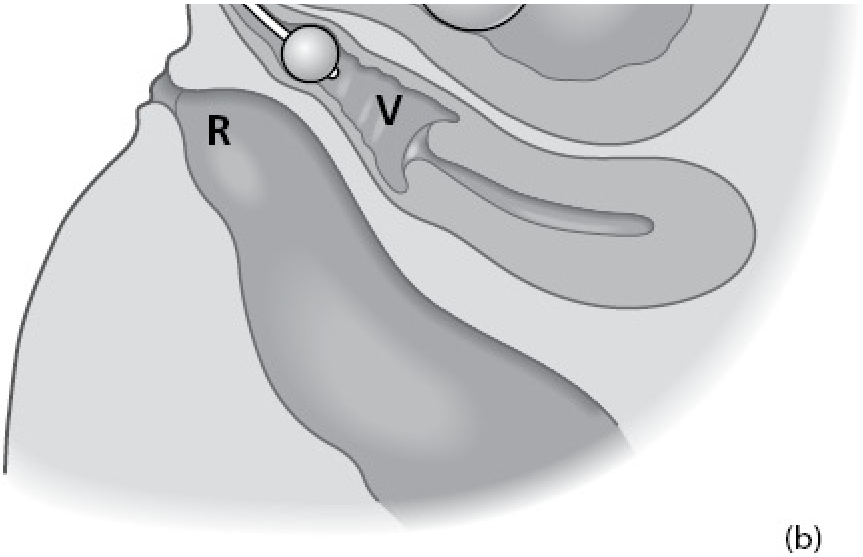
VAGINOPLASTY USING UROGENITAL MOBILIZATION

Total urogenital sinus mobilization (TUM) was described in 1997 by Alberto Peña as a technique to repair the UGS component of cloacal malformations. Currently, most surgeons are using this procedure for UGS repair, or a modification thereof made possible by the realization that the external sphincter of the urethra is spread along an anterior–posterior continuum and that there is a plane free of innervation along the dorsum of the urethra through which the UGS can be mobilized. TUM has the advantage of better visualization of the merging point and it obviates the need for vaginal separation, thus reducing the difficulty of the procedure and the likelihood of future structure of the orifice. The Fogarty balloon, placed in the vagina during the panendoscopy, allows the identification of the confluence (**Figure 80.12a**). In addition, in those cases where regained vaginal separation is required, it can be less extensive. In this technique, the mobilization of skin flaps is minimized because the confluence is brought closer to the perineum. The posterior dissection is similar to that done for a pull-through or flap procedures, with careful midline mobilization of the UGS off the rectum. The circumferential dissection in cases requiring a TUM is done past the pubourethral ligament, under the pubis, resulting in significant mobilization of the sinus.



(a)

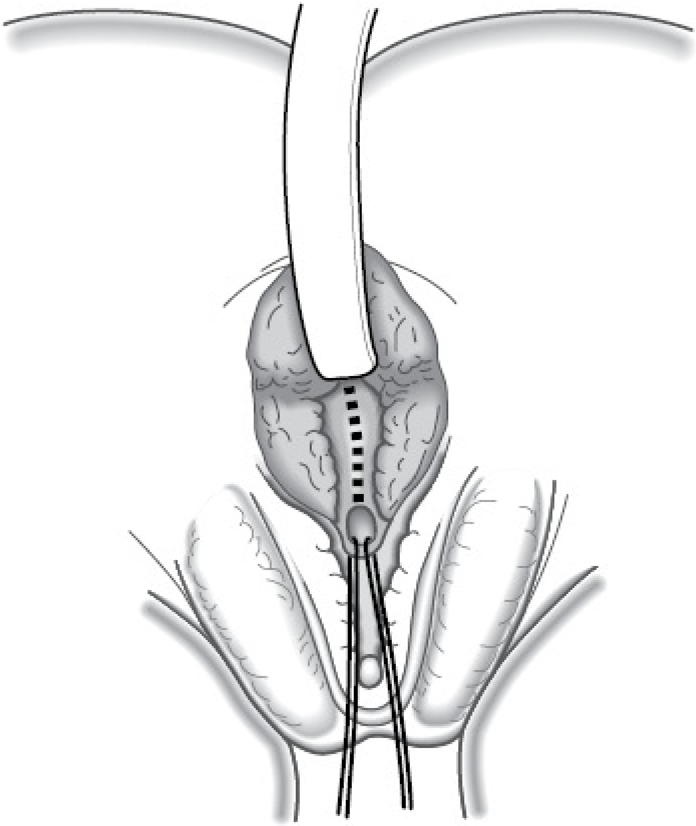




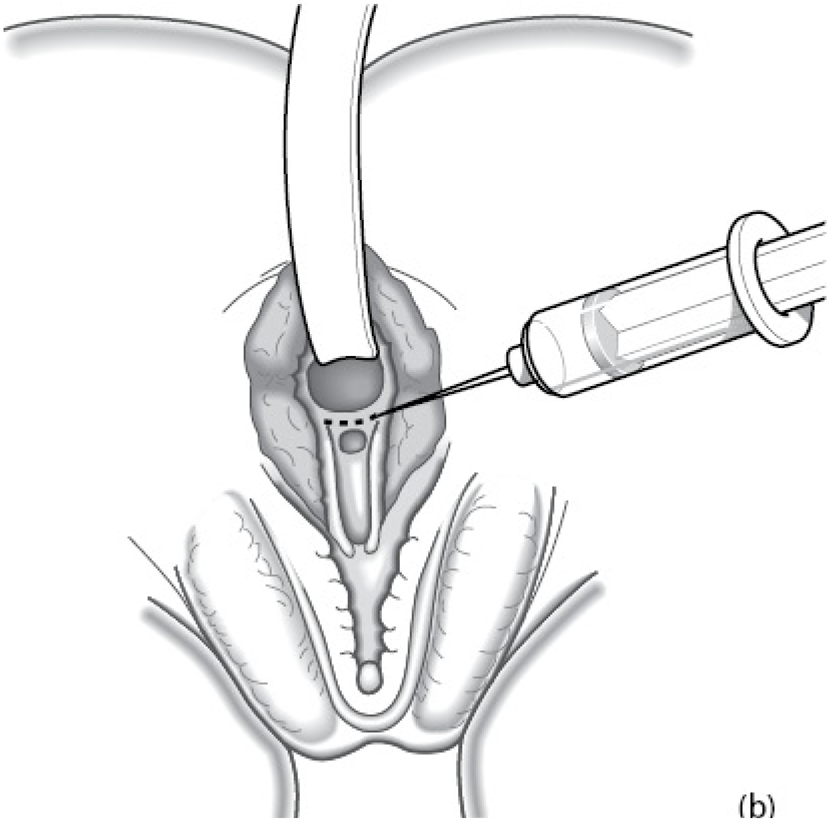
80.12a,b (a) High confluence; (b) low confluence.

In response to concerns for possible complications of urinary incontinence or prolapse, resulting from the dissection of the UGS beyond the pubourethral ligament, Rink and colleagues proposed in 2006 the use of a partial urogenital mobilization (PUM). In this technique, the anterior dissection stops at the pubourethral ligament, aiming to avoid compromising the innervation to the bladder outlet and clitoris (**Figure 80.12b**). Circumferential, partial mobilization of the UGS allows the mid-level vaginal confluence to be brought down to the perineum without tension, avoiding the need for separation of the vagina from the urethra as in prior pull-through vaginoplasties. This newer procedure is adequate in most cases except for patients with a very high confluence in whom additional mobilization past the pubourethral ligament may be needed.

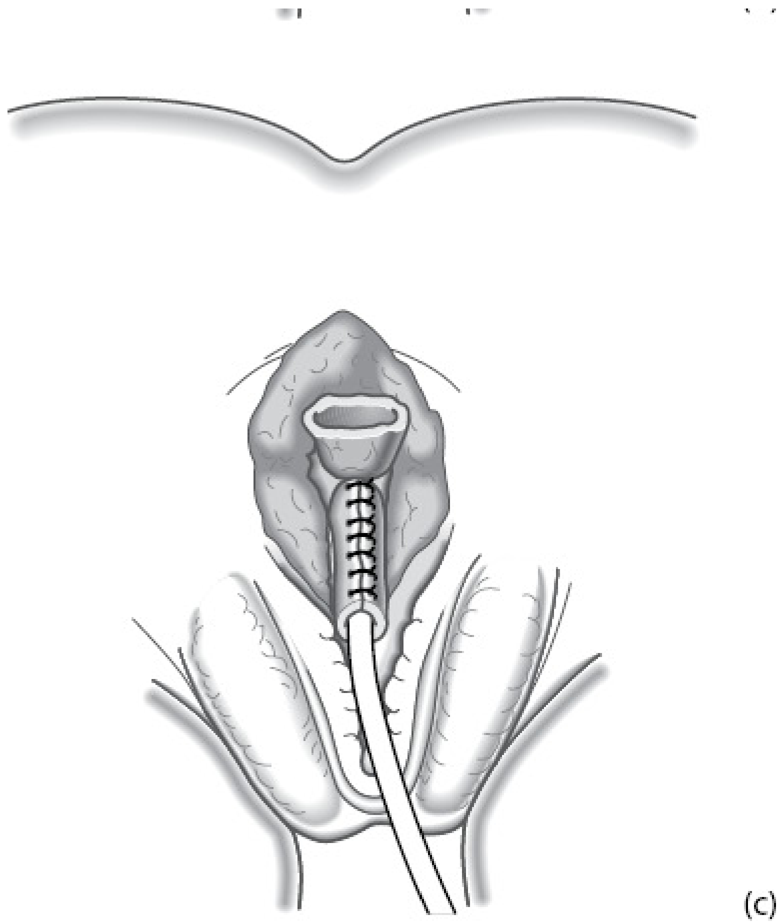
In high-confluence cases, if the perineum cannot be reached without tension, the vagina must be sharply dissected from the back wall of the bladder before the anastomosis to the inverted anterior perineal “U” flap can be attempted (**Figure 80.13a**). In these cases, the use of a prone position can facilitate the dissection of the vagina off the bladder.



(a)



(b)



80.13a–c

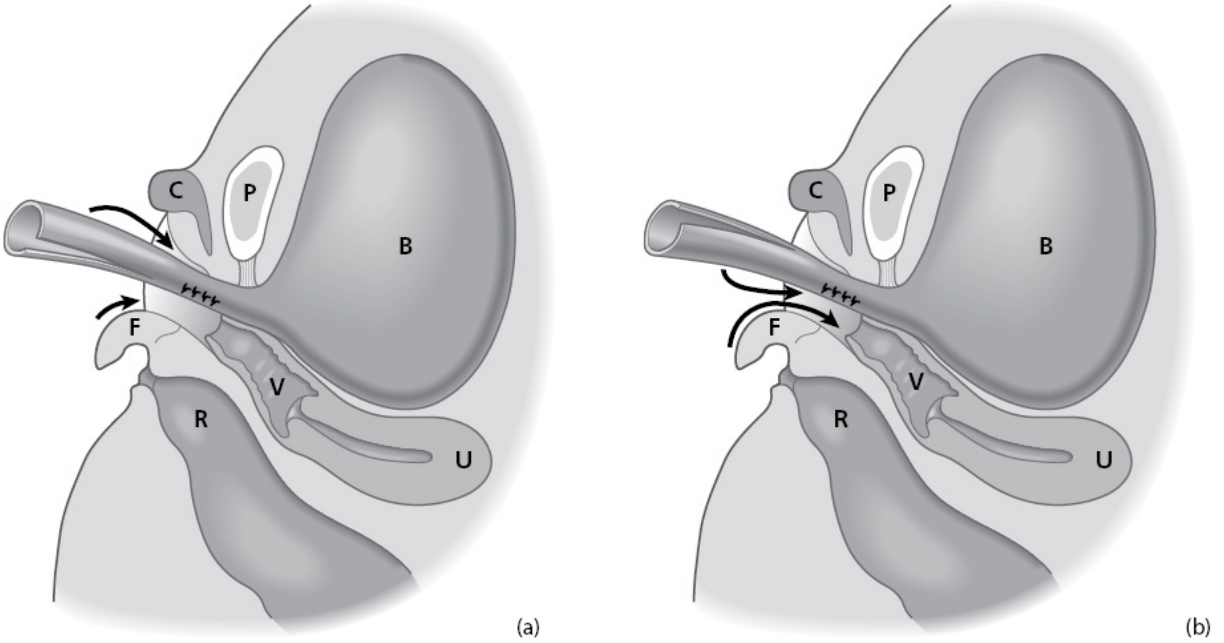
In both TUM and PUM, as previously described for low-confluence vaginoplasty, the distal segment of the vagina can be quite narrow, therefore its posterior wall must be incised up to normal caliber vagina to avoid a vaginal stricture (**Figure 80.13b**). The apex of the inverted perineal "U" flap is inserted into the apex of the vagina wall, and secured in position with interrupted full thickness sutures of 4/0 Vicryl™ (**Figure 80.13c**), as previously described for a low vaginoplasty (see **Figure 80.11a–c**).

SPLITTING THE UROGENITAL SINUS

The common UGS can be split to enhance the vaginoplasty in one of the following manners.

1. In very low-confluence cases, the authors do not mobilize the urogenital sinus, and incise it longitudinally on its ventral aspect down to the

confluence point; the lateral aspects of the opened sinus are then sutured to the medial aspects of the prepuce wings, thus resulting in a more normal anatomical configuration (**Figure 80.14a**).



80.14a,b

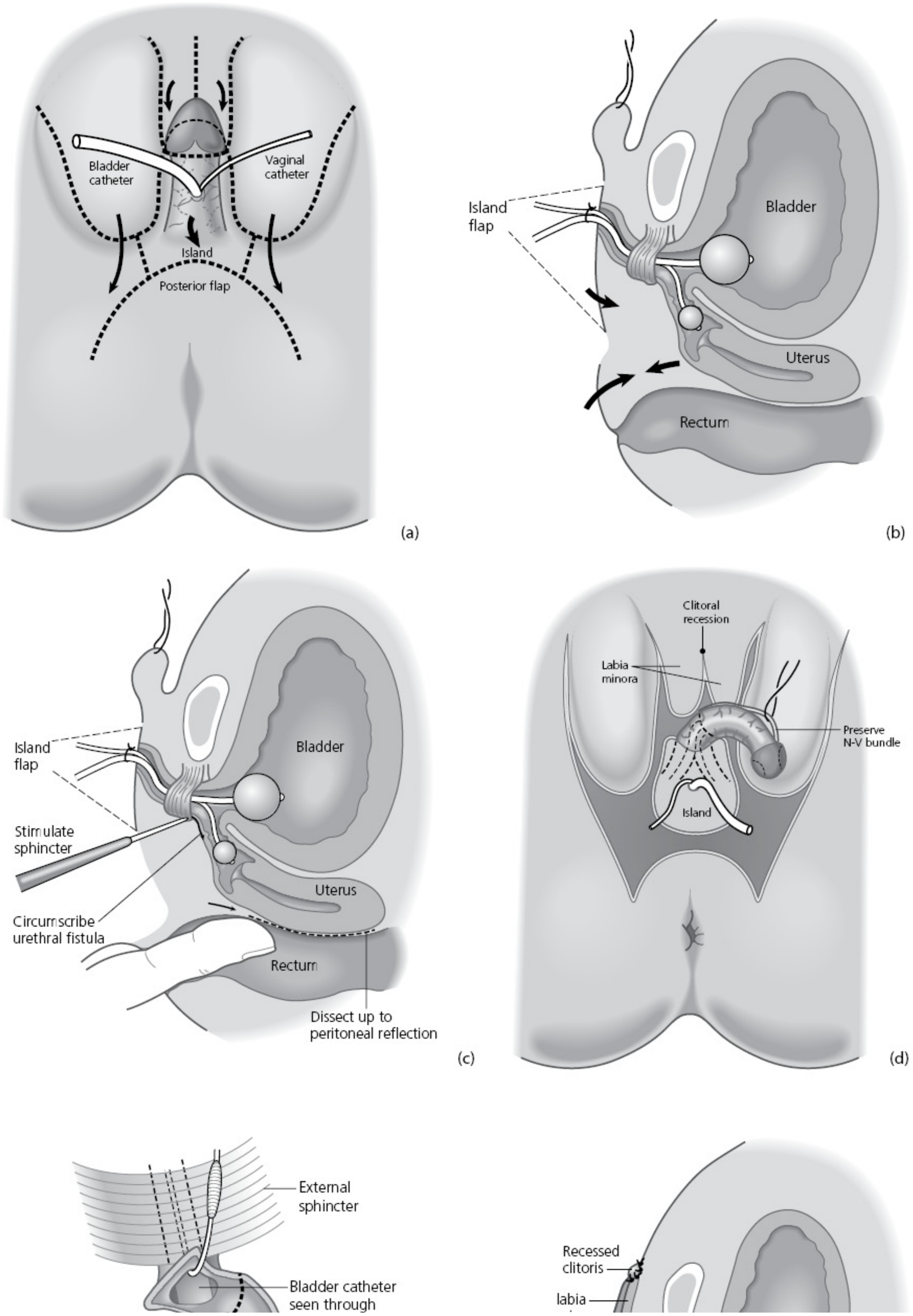
2. Split the sinus ventrally to fashion a mucosa-lined vestibule.
3. Split the sinus dorsally to create a flap which will enhance the anterior aspect of the vagina (**Figure 80.14b**).
4. Split the sinus laterally to create and rotate a flap to elongate the vagina.

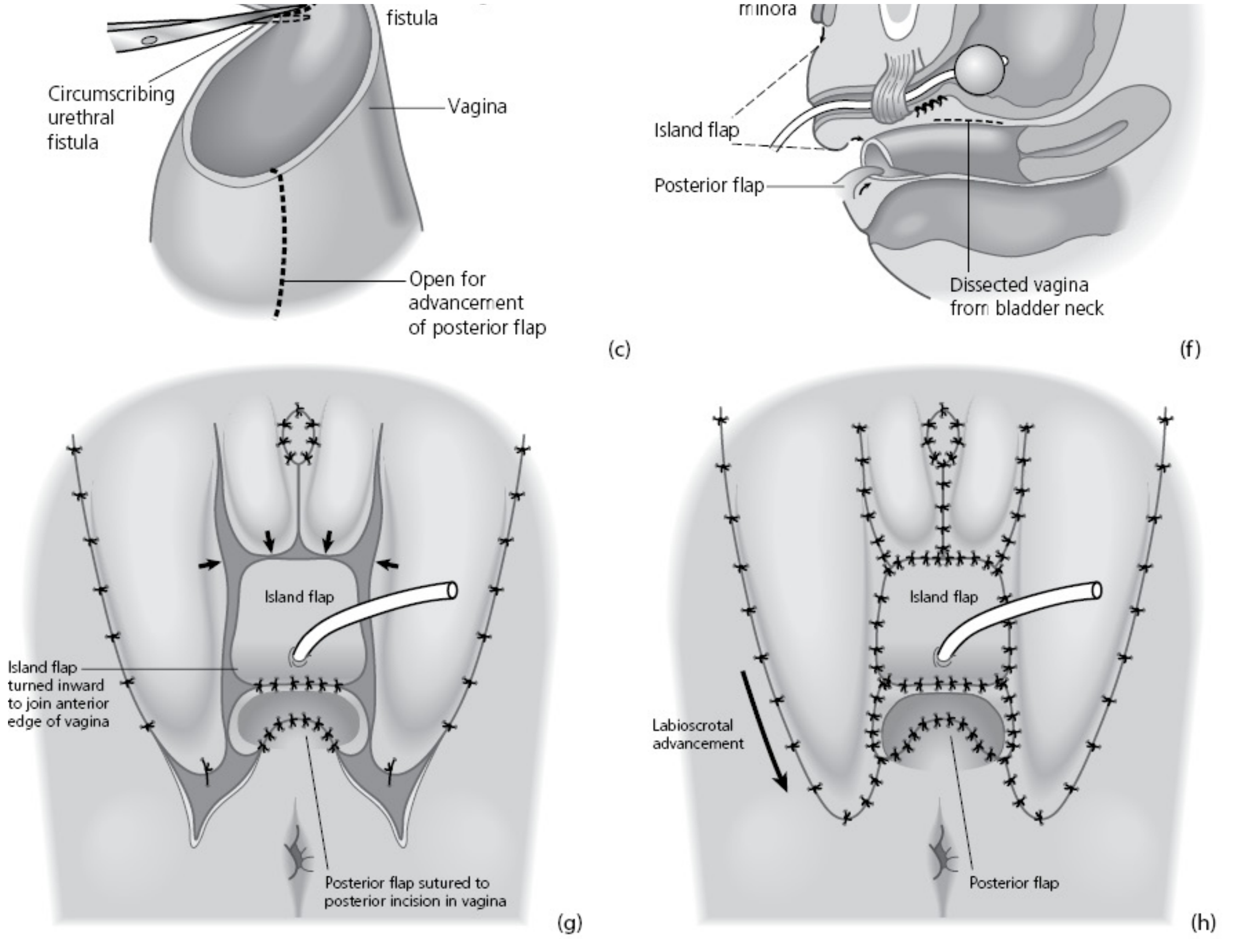
PULL-THROUGH VAGINOPLASTY FOR MID- AND HIGH-LEVEL VAGINAL CONFLUENCE

Before the advent of the UGS mobilization procedures, the majority of girls were reconstructed using a pull-through vaginoplasty; the UGS mobilization techniques are, however, now preferred for high, severely masculinized perineum, predominantly in cases of adrenogenital syndrome.

All patients undergo a total lower body preparation from nipples to toes, the legs are wrapped, and the lower body is passed through the aperture in the drapes, which allows the patient to be rotated either supine or prone during the procedure.

The initial aspects of the dissection for patients with moderate masculinization are similar to the previous description for lower confluences. The operation begins with a panendoscopy during which the confluence is localized, and a Fogarty catheter connected to a stopcock valve is maneuvered into the vagina, where the balloon is inflated and the valve closed. A Foley catheter is passed into the bladder (**Figure 80.15a**); both catheters are secured together with a silk suture (**Figure 80.15b**). An inverted perineal “U” flap is made, the UGS is exposed, and the balloon of the Fogarty catheter is palpated. Intermittent insertion of a finger inside the rectum, to flatten out its anterior wall, reassures the surgeon that the rectum has not been injured (**Figure 80.15c**). The vagina is mobilized circumferentially and meticulously separated off the bladder anteriorly. The UGS is closed with interrupted stitches of 6/0 PDS□. A labial flap or an anterior island flap, which for this procedure must be fashioned in the midline, can now easily reach the anterior vagina (**Figure 80.15d**). In patients with a very high confluence, the authors have found that rotating to the prone position improves visualization and surgical field, allowing the vagina to be safely mobilized off the UGS and bladder. The placement of a small malleable retractor into the vagina combined with slight upward traction facilitates the surgical dissection (**Figure 80.15e**). The opening in the UGS is closed with interrupted stitches of 6/0 PDS□. The vagina is then mobilized circumferentially and brought to the perineum, thus avoiding potential urethrovaginal fistula by overlapping suture lines over the UGS closure where the flap is sutured to the perineal skin flap as described for vaginoplasty (**Figure 80.15f–h**).


Bladder catheter
Vaginal catheter
Island
Posterior flap
(a)
Island flap
Bladder
Uterus
Rectum
(b)
Island flap
Bladder
Stimulate sphincter
Circumscribe urethral fistula
Uterus
Rectum
Dissect up to peritoneal reflection
(c)
Clitoral recession
Labia minora
Preserve N-V bundle
Island
(d)
External sphincter
Bladder catheter seen through
Recessed clitoris
labia


fistula
Circumscribing urethral fistula
Vagina
Open for advancement of posterior flap
(c)
minora
Island flap
Posterior flap
Dissected vagina from bladder neck
(f)
Island flap
Island flap turned inward to join anterior edge of vagina
Posterior flap sutured to posterior incision in vagina
(g)
Island flap
Labioscrotal advancement
Posterior flap
(h)

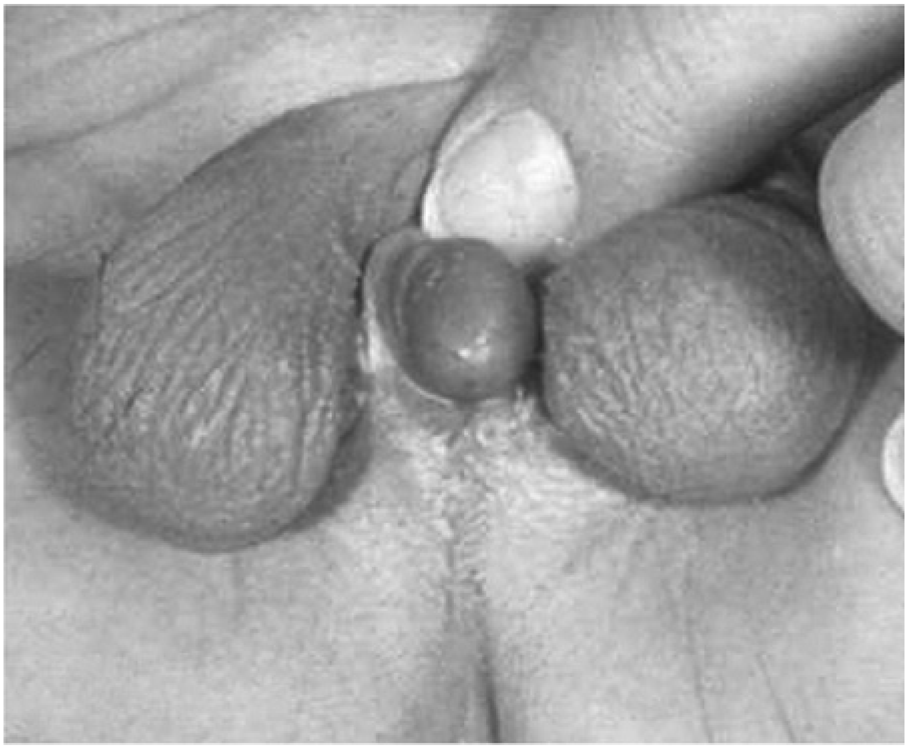
80.15a–h

Postoperative care of the female patient

A size 8 Fr Foley catheter is left indwelling for 3–4 days. Immobilization with a mermaid dressing, in which the lower extremities are wrapped with stockinette and an elastic bandage to prevent abduction, reduces tension on the suture lines. A soft foam or cotton is placed between the knees and ankles, and the anus is left exposed. This dressing is kept in place for 4–5 days. A broad-spectrum antibiotic is used during the first 2 postoperative days. Hormonal replacement appropriate to the patient's diagnosis is given.

RECONSTRUCTION FOR MALE GENDER ASSIGNMENT

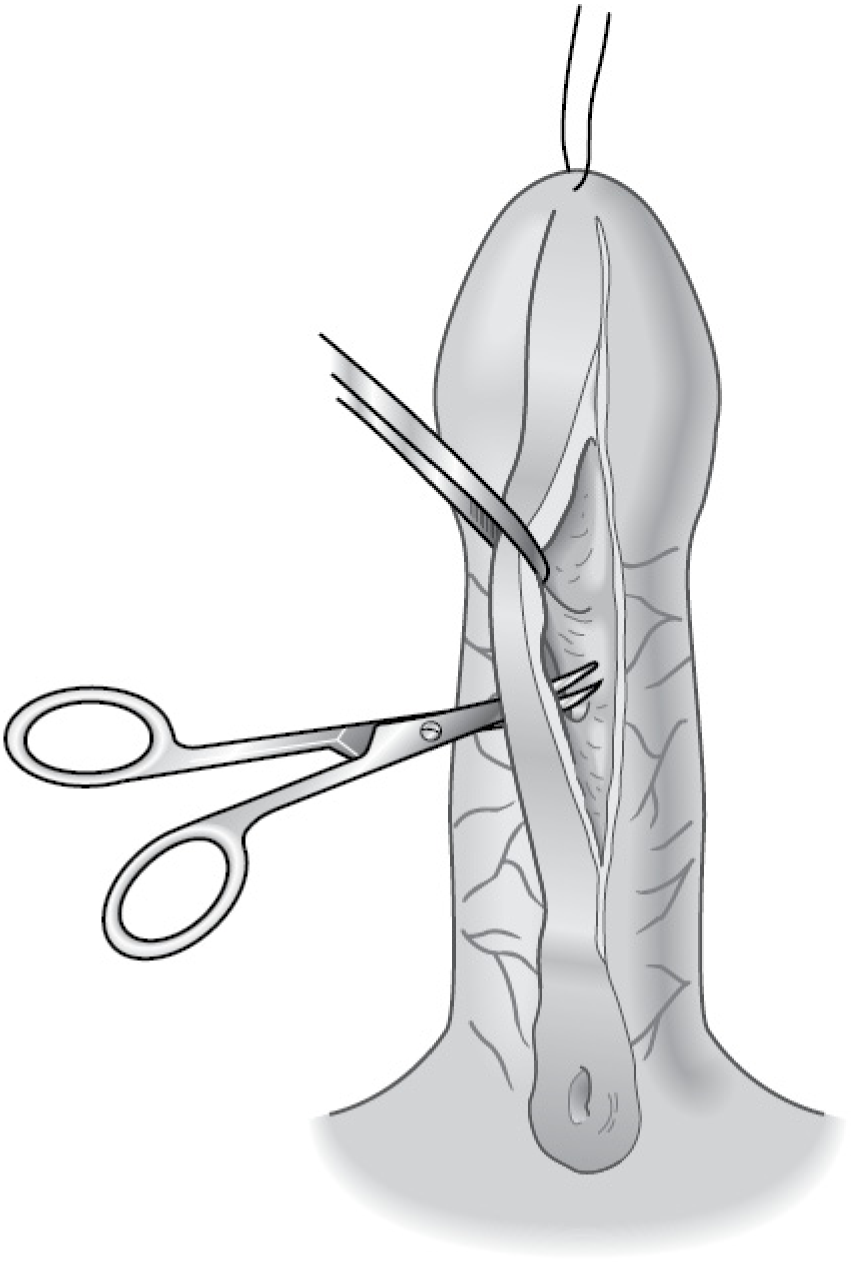
The treatment strategy is similar for all patients assigned to the male gender. Most of these patients have a small penis with a penoscrotal, scrotal, or a perineal hypospadias; a severe ventral curvature; and a partial or complete prepenile scrotum (**Figure 80.16**). Preoperative treatment with testosterone is helpful in those cases with a small penis and predicts salutary response at puberty.



80.16

Hypospadias repair

The authors' preferred approach is to perform a one-stage hypospadias repair, but, if necessary, the procedure can be staged. In patients with an adequate urethral plate, the tabularized incised plate urethroplasty is the preferred procedure. In this technique, the urethral plate is preserved; aggressive and meticulous degloving of the foreskin is carried out beyond the hypospadiac meatus into the scrotum, thus achieving in many cases significant correction of the ventral curvature (**Figure 80.17**). Then, an artificial erection is induced and, if needed, additional ventral penile curvature correction is accomplished with a dorsal plication, placing one or two 5/0 Prolene stitches at the 12 o’clock position dorsally, at the point of maximum angulation. In patients with a persistent, greater than 30-degree ventral penile curvature, the urethral plate can be dissected and elevated from the corpora cavernosa up to normal urethra, and, if needed, the correction can be enhanced with one to three transverse corporotomies, placing the first one at the point of maximal bending. The authors perform the urethroplasty in two layers, the first with interrupted subcuticular stitches of 7/0 PDS□ or 7/0 Vicryl™ with a TG-40 needle followed by a second running layer of the same material. The suture line should be covered with a well-vascularized dartos flap harvested from the dorsal prepuce, or with a tunica vaginalis flap.



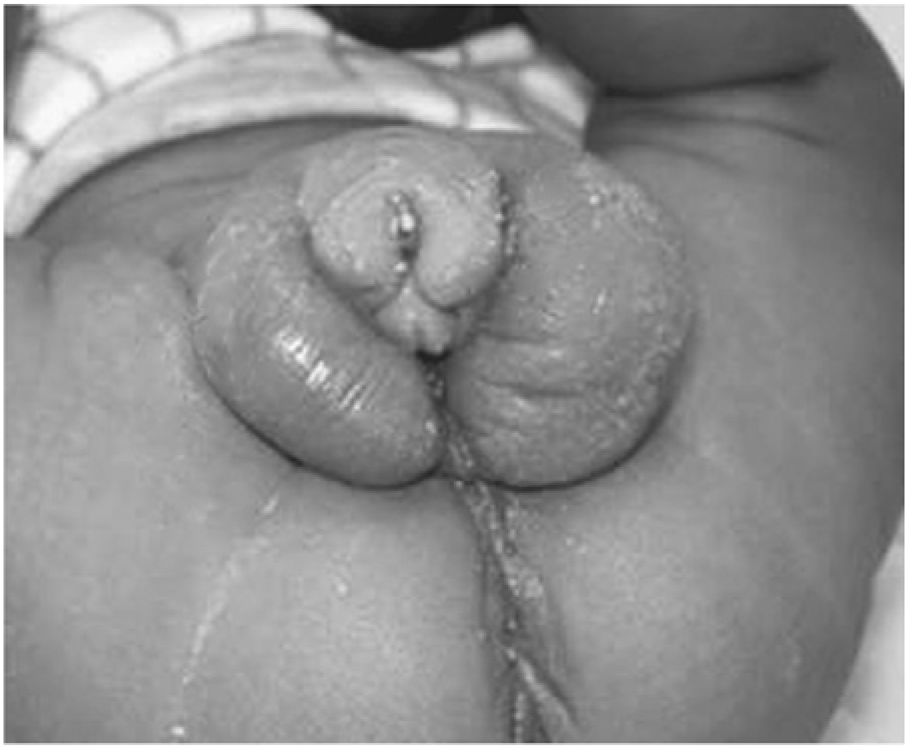
80.17

In cases requiring a multistage repair, a preputial graft or an oral mucosa graft can be used. If the penis has a severe ventral penile curvature, transverse dorsal corporotomies are made at the point of maximal concavity and a ventral dermal graft is sutured to the edges of the defect. The second stage is 6-9 months later and urethroplasty is performed. Two suture layers are used; the first is done with interrupted stitches of 7/0 PDS® or 7/0

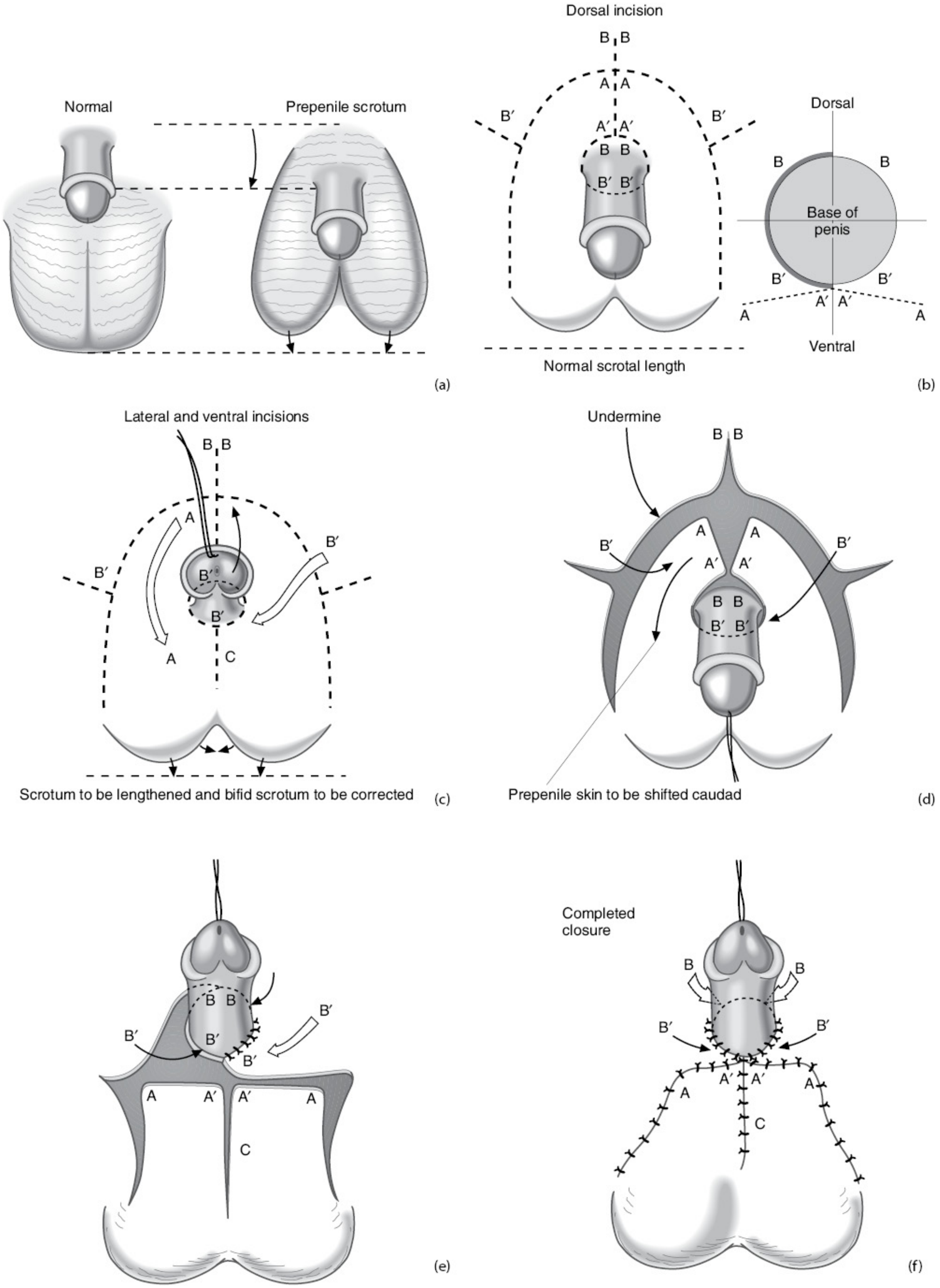
Vicryl™with a TG-40 needle, the second layer is a running suture of 7/0 PDS□. A penile dressing is fashioned with surgical wrapped around the penis; 2 × 2 gauze; coban tape; and Tegarderm, which minimizes the postoperative swelling and bleeding. The dressing is usually removed on the third postoperative day.

Penoscrotal transposition

Partial or complete penoscrotal transposition is often found in cases with penoscrotal and perineal hypospadiac (**Figure 80.18**). The least severe forms are known as bifid scrotum, prepenile scrotum (**Figure 80.19a**), and shawl scrotum. The scrotoplasty should be delayed until after the hypospadias repair is completed, to assure vascularization because the distal flaps needed for the hypospadias repair must be divided and displaced during correction of the prepenile scrotum. Six months or more should elapse between the urethroplasty and repositioning of the scrotum. This anomaly is repaired by displacing scrotal skin posteriorly and the penis anteriorly. The base of the penis is advanced forward onto the anterior abdominal wall by creating a square, distally based flap, which circumscribes the base of the penis. The flap is dropped distally to restore normal scrotal length. The abdominal wall is then undermined and swung around the base of the penis to join ventrally in the midline. It is important to mobilize the anterior abdominal wall flaps sufficiently so that midline separation does not occur. See **Figure 80.19b–f**.



80.18

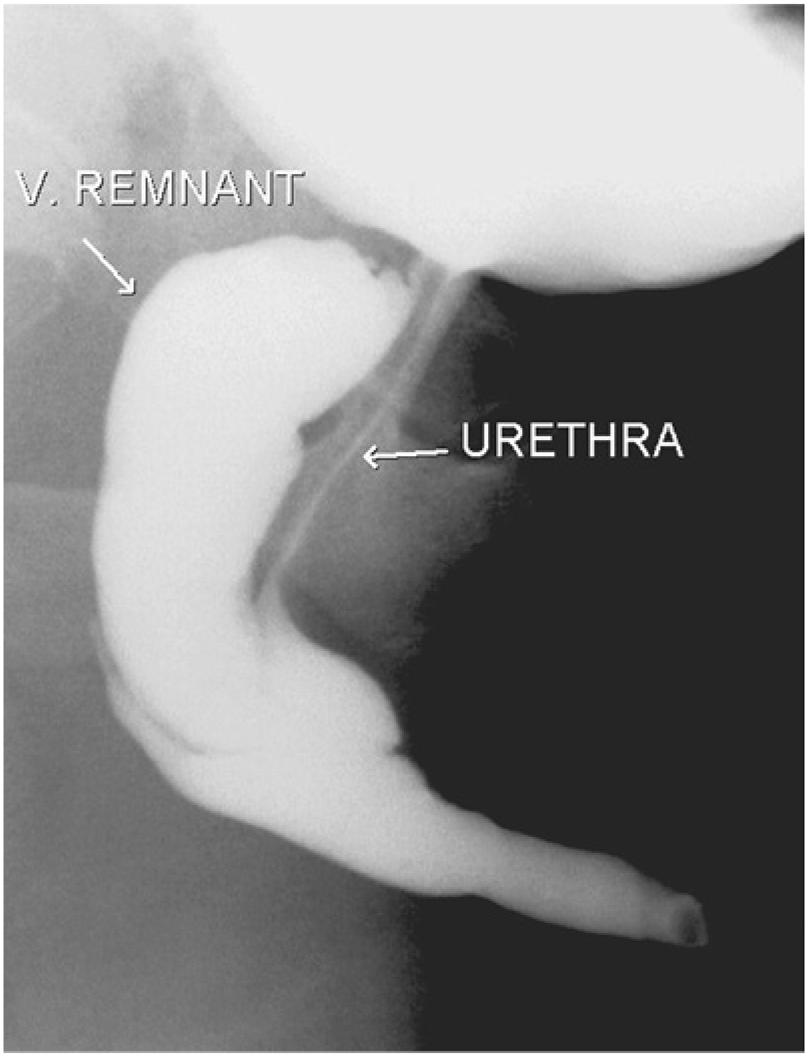

Normal
Prepenile scrotum
(a)
Dorsal incision
B B
A A
B′
B′
A′ A′
B B
B′ B′
Normal scrotal length
Dorsal
B
B
Base of penis
B′
B′
A′ A′
A
A
Ventral
(b)
Lateral and ventral incisions
B B
A
B′
B′
B′
B′
A
C
Scrotum to be lengthened and bifid scrotum to be corrected
(c)
Undermine
B B
A A
B′
B′
A′ A′
B B
B′ B′
Prepenile skin to be shifted caudad
(d)
B B
B′
B′
B′
B′
A A′ A′ A
C
(e)
Completed closure
B
B
B′
B′
A′ A′
A
A
C
(f)

80.19a–f

Removal of retained Müllerian ducts and creation of neoseminal vesicle

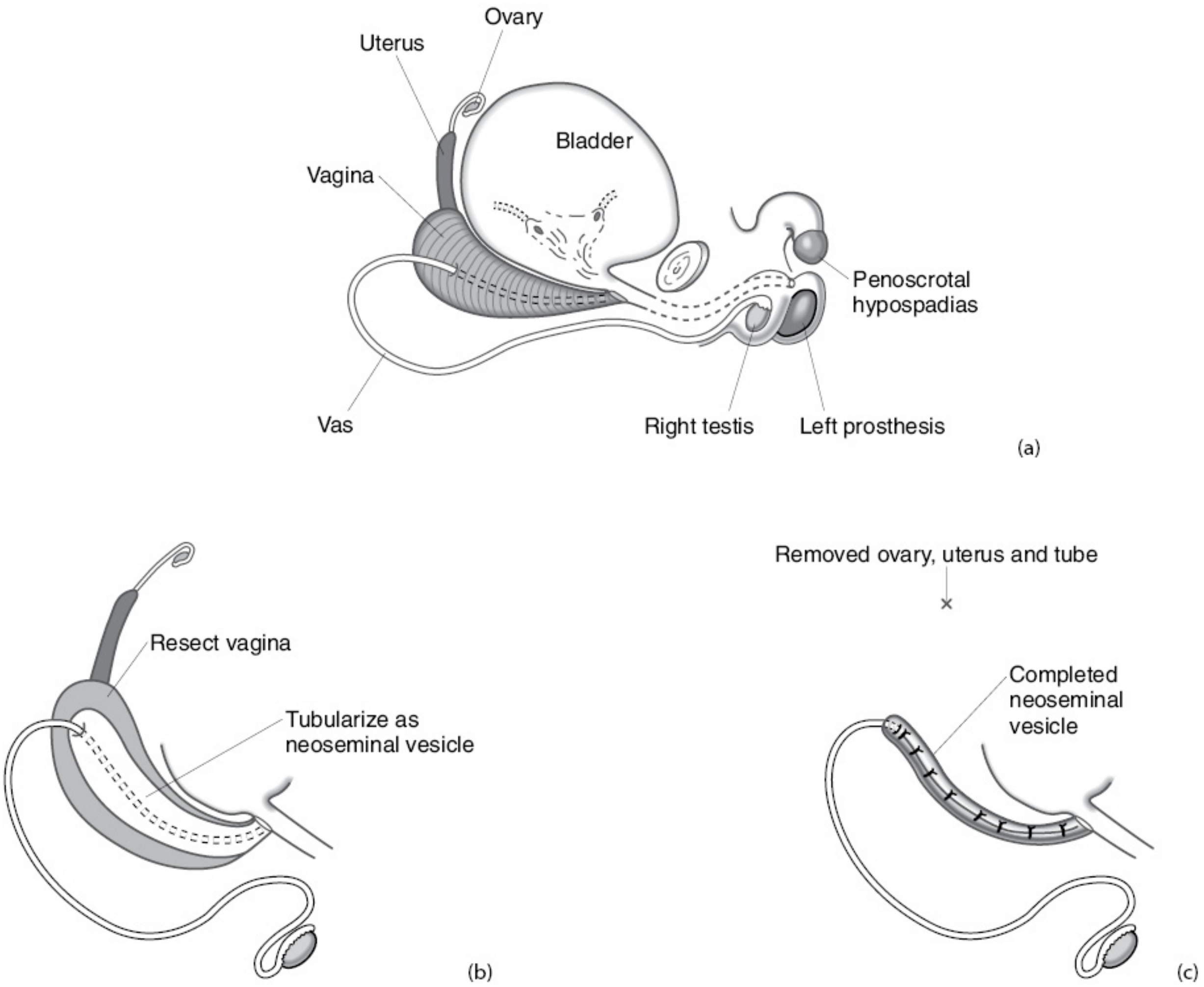
In patients with severe hypospadias, ovotesticular disease, or MGD who have been assigned the male gender, the retained Müllerian ducts can become quite enlarged leading to recurrent urinary tract infections, epididymo-orchitis, urinary retention, secondary incontinence due to urine trapping, and infertility. Malignant transformation has also been reported. In cases with these complications, the Müllerian duct remnant may have to be removed.

Surgical treatment of Müllerian duct remnants is challenging, because of their close proximity to the ejaculatory ducts, pelvic nerves, rectum, vas deferens, and ureters. A number of surgical approaches have been described, including transperitoneal, posterior with rectal retraction, anterior and posterior saggital transrectal, transtrigonal, perineal, transurethral fulguration, laparoscopic, and robotic. The authors have successfully used the transtrigonal technique; however, the laparoscopic or robotic-assisted procedures are less invasive and minimize possible damage to adjacent anatomical structures. The case corresponding to **Figure 80.20** was successfully operated by a robotic-assisted laparoscopic procedure.



80.20

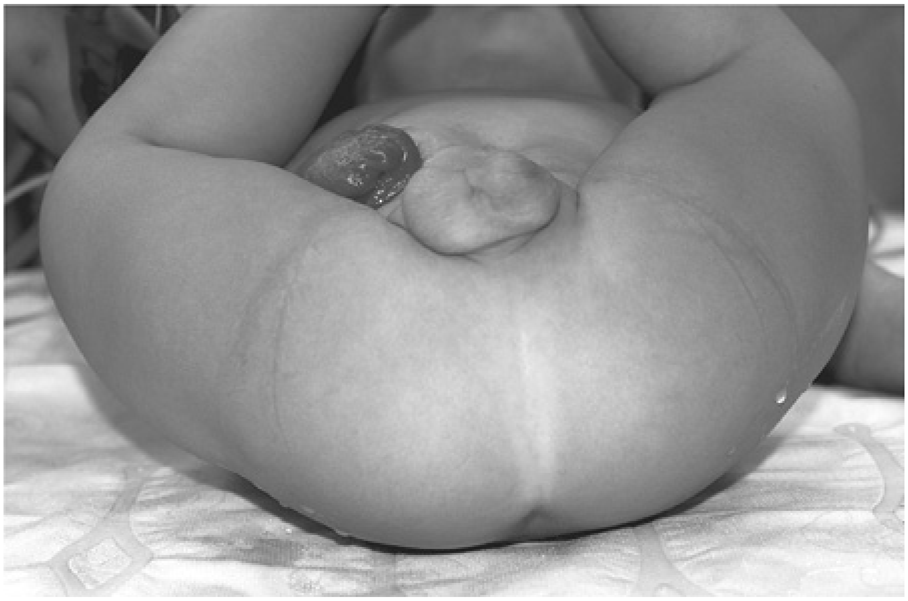
In males with retained Müllerian ducts, the vas deferens often lies within the sidewall of the dilated vagina (**Figure 80.21a**); its course, however, cannot usually be palpated in the thickened vaginal wall, therefore a strip of vagina surrounding the predicted course of the vas from its entrance to its termination with the urethra is preserved while the uterus and the remainder of the vagina are resected (**Figure 80.21b**). The vaginal strip is turned in and tubularized from the proximal vas to the point of union near the urethra using interrupted sutures of 5/0 Vicryl□ (**Figure 80.21c**). If the vas courses in the wall of the uterus, it is not possible to perform this procedure.


Ovary
Uterus
Bladder
Vagina
Penoscrotal hypospadias
Vas
Right testis
Left prosthesis
(a)
Resect vagina
Tubularize as neoseminal vesicle
(b)
Removed ovary, uterus and tube
Completed neoseminal vesicle
(c)

80.21a–c

Penile agenesis

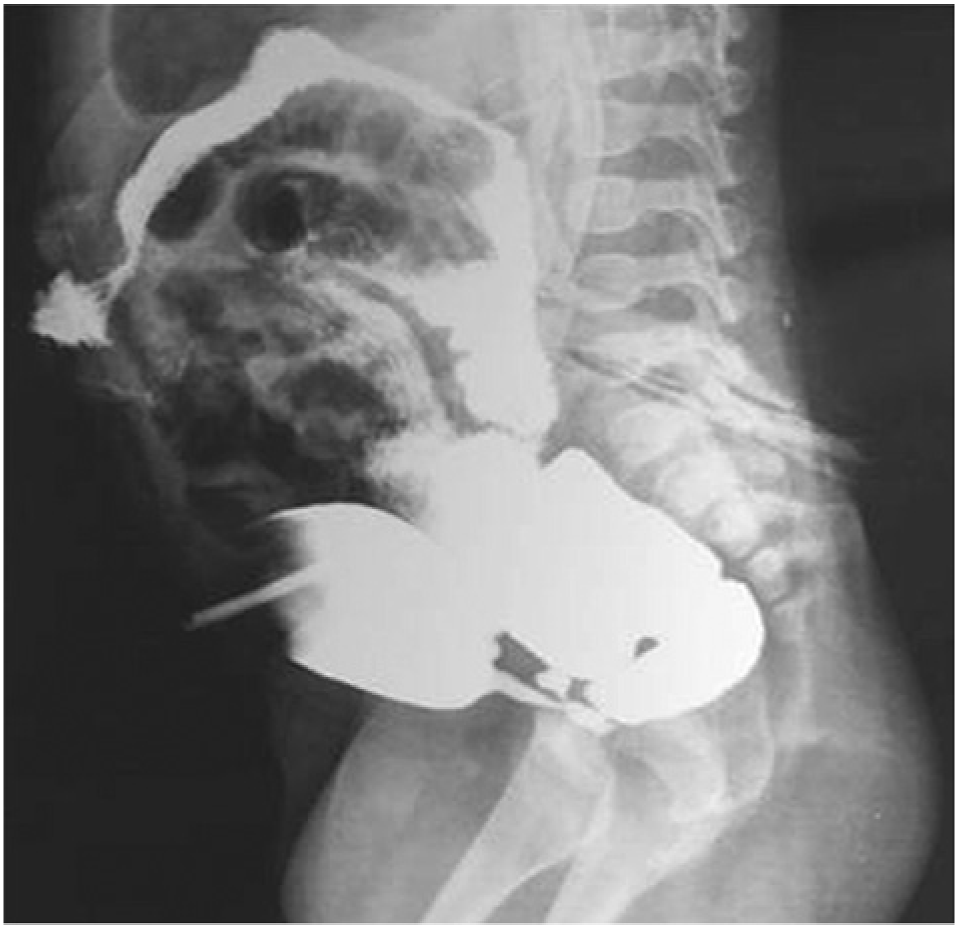
Penile agenesis occurs in 1 in 30 million births. It appears to be the result of a development failure of the genital tubercle during the fourth week of embryogenesis. The scrotum appears normal and contains normal testicles (**Figure 80.22**). Patients can present with an imperforate anus and a rectourethral fistula, with a normal anus and a rectourethral fistula, or with the urethra located in the perineum inside a skin tag resembling a foreskin. Patients are otherwise normal 46,XY males.



80.22

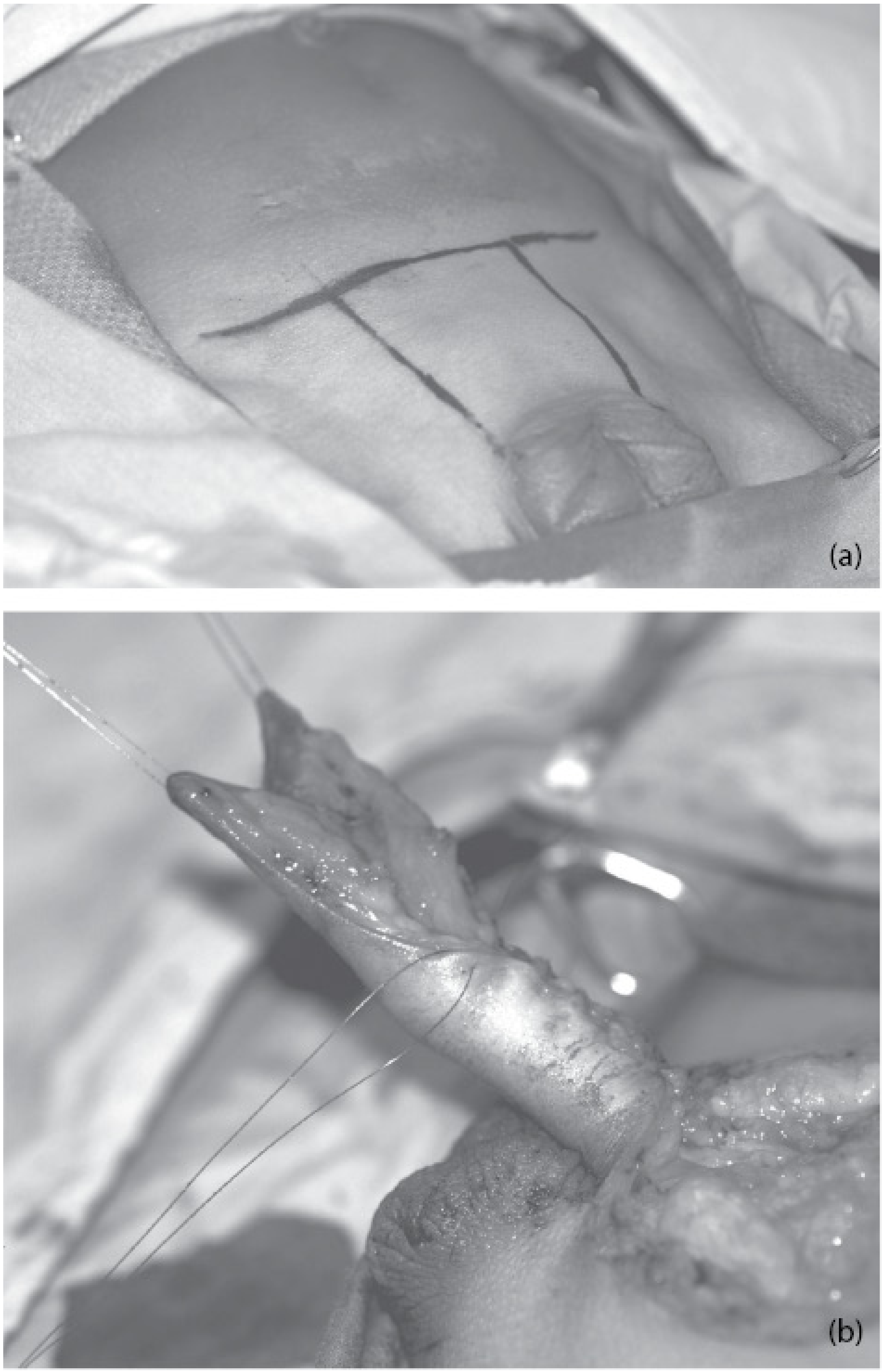
In the past, gender reassignment was the most frequent choice for these patients. Initial surgical treatment of female-assigned patients requires bilateral orchiectomy. Cases with an associated imperforate anus and rectourethral fistula need an urgent colostomy and a vesicostomy. Children assigned to the female sex require the creation of a neovagina, which is most frequently performed with a sleeve of sigmoid colon. However, some patients have been unhappy with the assigned female sex, and prefer to be males.

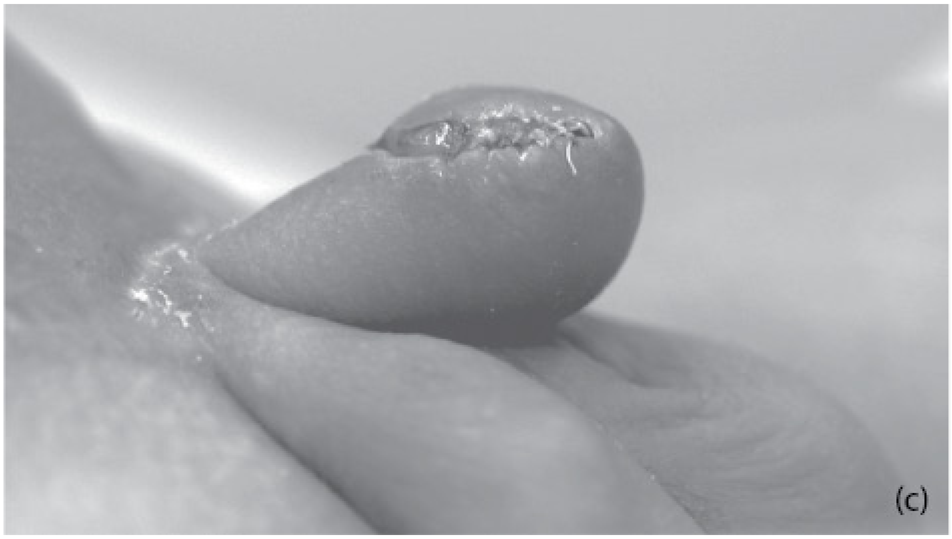
The diagnostic evaluation of patients with penile agenesis includes a renal ultrasound (US), pelvic MRI, retrograde urethrogram, and, in cases associated with an imperforate anus, a distal colonogram through the mucous fistula, using hydrosoluble contrast material (**Figure 80.23**). The colonogram can be combined with an antegrade VCUG via the cutaneous vesicostomy. An experienced medical team must evaluate newborns with penile agenesis. Families must be given all available information regarding sex assignment, surgical procedures, and immediate and long-term results so that they can make a decision which is in the best interest of their child. Genetic evaluation with whole genomic sequencing is recommended for genetic counseling and for understanding etiology.



80.23

In adolescents and adults, the most frequently used phalloplasty procedure is the radial forearm flap, which is a complex and rare operation, performed only in highly specialized centers. The description by De Castro of a phalloplasty technique and complete urethroplasty using a quadrangular lower abdominal flap can bridge the interval between childhood and adolescence until a more definitive procedure can be performed (**Figure 80.24a–c**). The quadrangle of lower abdominal flap fashioned to create the new penis is 4 cm × 5 cm for babies, and slightly larger in an older child. De Castro recommends the use of oral or bladder mucosa for the urethroplasty, although the single-stage buccal mucosa urethroplasty has had a high complication rate.


(a)
(b)



80.24a–c

The authors are in favor of staging the surgical reconstruction. As mentioned before, cases with an imperforate anus and a rectourethral fistula require an urgent colostomy with a mucous fistula and a vesicostomy, followed a few months later by a posterior sagittal rectourethral pull-through, with subsequent closure of both the vesicostomy and colostomy. Patients born with a normal anus and a rectourethral fistula can be operated through a transperineal approach using a hyperextended position. The authors have used the quadrangle lower abdominal flap to fashion the neopenis in three patients, but prefer a two-stage buccal mucosa urethroplasty to avoid or minimize complications.

Vaginal agenesis

Vaginal agenesis is known as Mayer–Rokitansky–Kuster–Hauser syndrome. It is a rare abnormality affecting approximately 1 in 5000–10 000 females. In this condition, there is absence of the proximal portion of the vagina, resulting from failure of the sinovaginal bulbs to develop and form the vaginal plate.

This condition can be discovered at birth when the examination does not reveal a vaginal opening, but most cases present with primary amenorrhea.

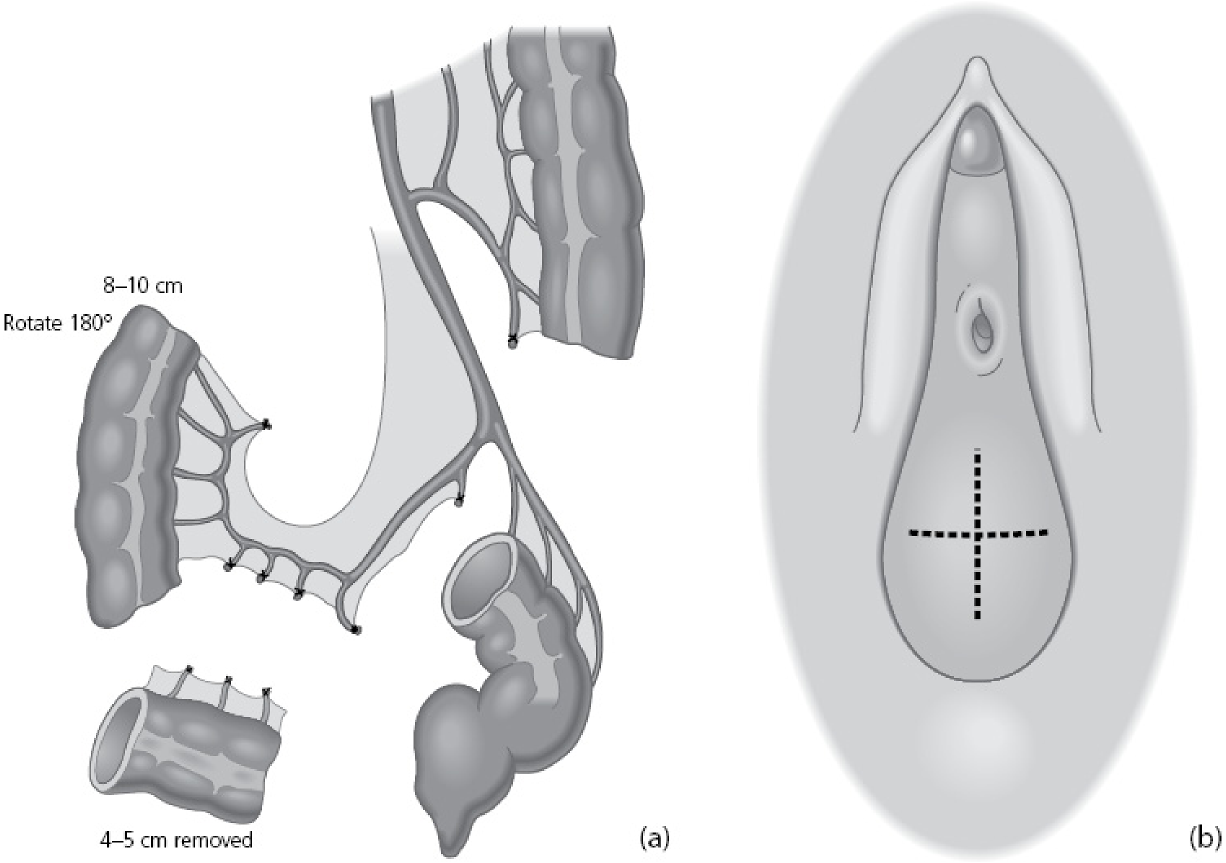
A small group of patients with imperforate anus will have vaginal agenesis, and each of the females requires inspection of the vaginal orifice prior to proceeding with definitive pull-through. Few patients complain of dyspareunia or failed intercourse. Physical examination reveals an absent vagina, but the hymen and a distal vaginal dimple or even an introitus are present, because these structures are derived from the urogenital sinus. The diagnosis can be confirmed by pelvic US and MRI.

NON-SURGICAL TREATMENT

Vaginal dilatation is an alternative to surgery in patients with a generous vaginal dimple or introitus. A program of daily, graduated, dilatations over several months, using Hegar or similar sounds can result in a good-size vagina to allow intercourse. Once a satisfactory vaginal size is obtained, regular sexual intercourse maintains an adequate vaginal cavity.

SURGICAL TREATMENT

Different techniques for vaginal construction in patients with vaginal agenesis have been used, including the construction of a skin neovagina, and the creation of an intestinal neovagina using sigmoid, cecum, and small intestine. Our preferred technique for vaginal replacement is the use of an 8–10 cm segment of distal sigmoid colon based on the left colic or superior hemorrhoidal vessels (**Figure 80.25a**). Mechanical and antibiotic bowel preparation must be done before the procedure. A lower transverse abdominal incision gives adequate exposure. A supine position with legs spread and the knees slightly bent, using Allen stirrups, is recommended. The sleeve of sigmoid is selected between non-crushing clamps; a short distal sigmoid segment can be discarded to increase the length on the mesenteric vasculature for the neovagina. The sigmoid sleeve can be rotated 180 degrees to allow placement in the perineum. The proximal end is closed with two layers of absorbable suture material. Either a cruciate or H-shaped large perineal opening is made (**Figure 80.25b**); the rectovesical space is dissected to create a large tunnel that will allow the easy introduction of two fingers in the perineum. The sigmoid segment is pulled through the perineal channel and anastomosed directly to the perineum with absorbable sutures. Two-point fixation between the proximal end of the neovagina and the presacral fascia prevents prolapse of the bowel. The authors do not elevate the proximal vaginal dimple into the cul-de-sac to perform the anastomosis because, in our experience, it results in a higher frequency of strictures. Alternatively, a laparoscopic-assisted procedure can be done with a satisfactory outcome.



80.25a,b

The creation of a sigmoid neovagina has several advantages, such as natural lubrication without excessive mucous production, which can be avoided by reducing the diameter of the pull-through. Long-term use of a stent is avoided, stenosis is infrequent, and most patients have normal sexual intercourse.

SUMMARY

The surgical treatment of DSD should be undertaken by experienced surgeons who must be familiar with all the available techniques, so satisfactory repairs can be performed either to support the male or the female gender. They must also be familiar with the developmental biology and genetics of the urogenital and external genital organs, as well as have an armamentarium of surgical techniques allowing them to vary procedures according to the needs of each patient. The treatment plan must be thoroughly discussed with parents, with the goal of giving the child the most satisfactory quality of life possible. Long-term studies are, however, needed to assess the functional and sensory outcomes of newer surgical techniques.

ACKNOWLEDGMENTS

The authors would like to acknowledge that they received written authorization from Elsevier to reproduce the following figures from Chapter 123 (Disorders of sexual development, by Rafael V. Pieretti and Patricia K. Donahoe) in *Pediatric Surgery* (2011): Figures 123.4a,b; 123.6a,b; 123.7; 123.8; 123.9; 123.10a,b; 123.11; 123.12; 123.13; 123.14a–c; 123.15a,b; 123.16a–c; 123.17a–h; 123.19; 123.22a–c; 123.24; 123.25a–c; 123.26a,b; 123.27. In addition, the authors obtained authorization to use, modify, and adapt some paragraphs from the basic science and the surgical treatment sections of Chapter 123.

FURTHER READING

Baskin LS, Erol A, Li YW, et al. Anatomical studies of the human clitoris. *J Urol* 1999; 162: 1015–20.

De Castro R, Merlini E, Rigamonti W, Macedo A Jr. Phalloplasty and urethroplasty in children with penile agenesis: Preliminary report. *J Urol* 2007; 177: 1112–16.

Donahoe PK, Schnitzer JJ, Pieretti RV. Ambiguous genitalia in the newborn. In: O'Neill JA, Rowe ML, Grosfeld JL et al. (eds). *Pediatric Surgery*, 6th ed. St Louis: Mosby-Year Book; 2005: pp. 1911–34.

Ludwikowski B, Oesch Hayward I, Gonzalez R. Total urogenital sinus mobilization: Expanded applications. *BJU Int* 1999; 83: 820–2.

MacLaughlin DT, Donahoe PK. Sex determination and differentiation. *N Engl J Med* 2004; 350: 367–78.

Peña A. Total urogenital mobilization – an easier way to repair cloacas. *J Pediatr Surg* 1997; 32: 263–7.

Rink RC, Metcalfe PD, Kaefer MA, et al. Partial urogenital mobilization: A limited proximal dissection. *J Pediatr Urol* 2006; 2: 351–6.

Rink RC, Szymanski KM. Disorders of sexual development: Surgical management. In: Lima M, Manzoni G (eds). *Pediatric Urology: Contemporary Strategies from Fetal Life to Adolescence*. New York: Springer; 2014: pp. 247–58.

81

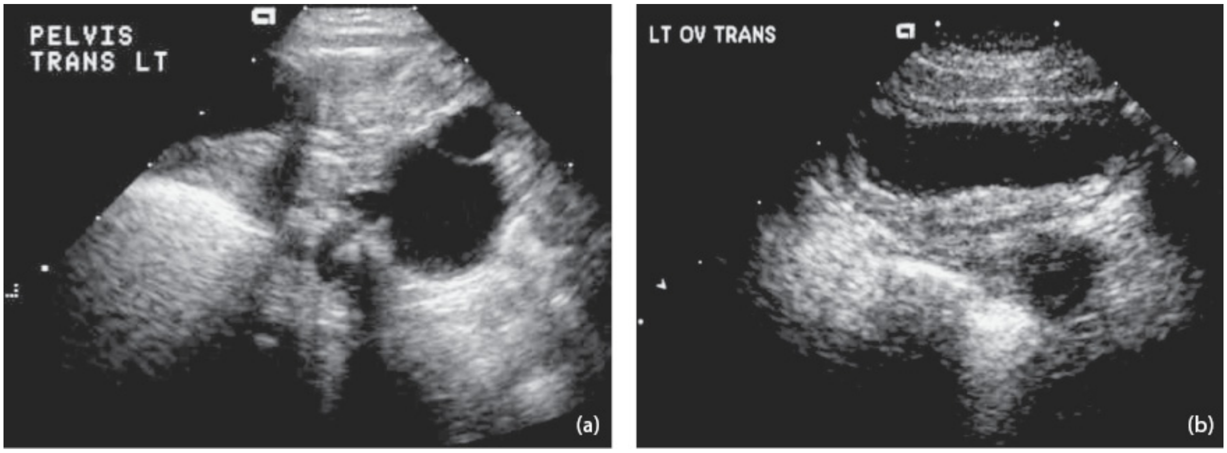
Ovarian cyst and tumors

BRYAN J. DICKEN AND DEBORAH F. BILLMIRE

PRINCIPLES

Ovarian cysts are relatively common and should be regarded as a separate entity from cystic ovarian neoplasms when considering surgical management. Follicular cysts are not neoplastic and are sensitive to hormonal stimulation. For clarity, ovarian cysts and neoplasms will be discussed separately.

Complex ovarian cysts have undergone hemorrhage and are heterogeneous with internal septa and fluid-debris levels (**Figure 81.1a and b**). Most are asymptomatic and resolve spontaneously with preserved ovarian function. As with simple cysts, surgical treatment should emphasize preservation of ovarian parenchyma. Open or laparoscopic detorsion, if present, with unroofing or fenestration of the cyst wall is indicated.



81.1a,b

NON-NEOPLASTIC CYSTS

Neonatal ovarian cysts

Neonatal ovarian cysts arise from maternal estrogen or human gonadotropin. The majority are of no clinical significance and will resolve spontaneously within 3–4 months. The risks of torsion and/or hemorrhage cannot be predicted on the basis of size.

Simple cysts are completely anechoic with a thin wall on ultrasound (US). They may be followed with serial US unless symptomatic due to large size. Cyst aspiration is associated with high recurrence rates and carries the risk of inducing hemorrhage. Surgical management by open or laparoscopic technique is preferred for symptomatic cases, and consists of fenestration or unroofing.

Ovarian cysts beyond the neonatal period

Beyond the neonatal period, follicular cysts are detected most often after puberty.

Females presenting with precocious puberty, vaginal bleeding, and an ovarian cyst should prompt a pediatric endocrinology consultation to exclude McCune–Albright syndrome. This is a sporadic disorder associated with autonomous estrogen-producing ovarian cysts that resolve spontaneously. Children usually present between 1 and 5 years of age with vaginal bleeding. This disorder is most frequently confused with an ovarian tumor, such as a juvenile granulosa cell tumor, resulting in unnecessary surgery.

Follicular cysts may be asymptomatic or present with pain due to large size, hemorrhage, or torsion. Most simple cysts will resolve spontaneously within three or four menstrual cycles. Indications for surgical intervention include persistent symptoms, pain, or evidence of torsion. As with neonatal non-neoplastic cysts, emphasis should be placed on ovarian parenchyma-preserving procedures.

Surgery for non-neoplastic ovarian cysts

If laparotomy is preferred, a low transverse incision on the side of the lesion is convenient (**Figure 81.2**). The cyst is delivered through the incision. A tense cyst may be needle-aspirated to facilitate grasping and delivery through a smaller incision. The ovary is inspected to locate the normal parenchyma and pedicle. The cyst may be enucleated if a plane is easily developed without injury to the ovarian parenchyma. A laparoscopic approach is an acceptable alternative, with 3 mm working ports, and a 5 mm umbilical port (may be upsized to a 10 mm if necessary) to facilitate removal of the cyst or cyst wall.

81.2

Alternatively, the free margin of the cyst wall is excised with cautery. A portion of the cyst wall should be sent for pathologic evaluation. The ovary is returned to the peritoneal cavity after confirming hemostasis. The fascia is closed, followed by subcuticular skin closure.

Preoperative assessment

US is the preferred initial modality for characterizing ovarian lesions. Simple cysts can be distinguished from neoplastic cysts and blood flow to the ovary can be determined. Serial examinations can be done for asymptomatic cysts. With precocious puberty, estrogen levels should be measured. US findings suggestive of neoplasm require further workup, as described within the ovarian neoplasm section (Table 81.1).

Table 81.1 **Summary of management of ovarian cysts and tumors**

	Pre-operative action	Surgery	Action points
Non-neoplastic			
Congenital cyst	Serial US	Laparoscopy or laparotomy	Most cysts resolve without intervention. Surgery for non-resolving/symptomatic
Simple cyst	US, tumor markers	Laparoscopy or laparotomy	Consult endocrinology if precocious puberty. Follow-up US in 6–8 weeks
Torsion of ovary	US, tumor markers	Laparoscopy or laparotomy	US follow-up in 6–8 weeks
Neoplastic			
Ovarian tumor	US, CT/MRI, tumor markers	Laparoscopy (Stage 1, <10 cm)	Annual US + tumor markers. Complete staging mandatory in all cases[a]
		Laparotomy (preferred)	Enucleation if solid/mixed and negative tumor markers is acceptable

[a] Peritoneal fluid for cytology, evaluation of peritoneal surfaces (biopsy suspect lesions), omentum (resect if abnormal) and retroperitoneal lymph nodes evaluation (biopsy abnormal nodes), inspection of contralateral ovary.

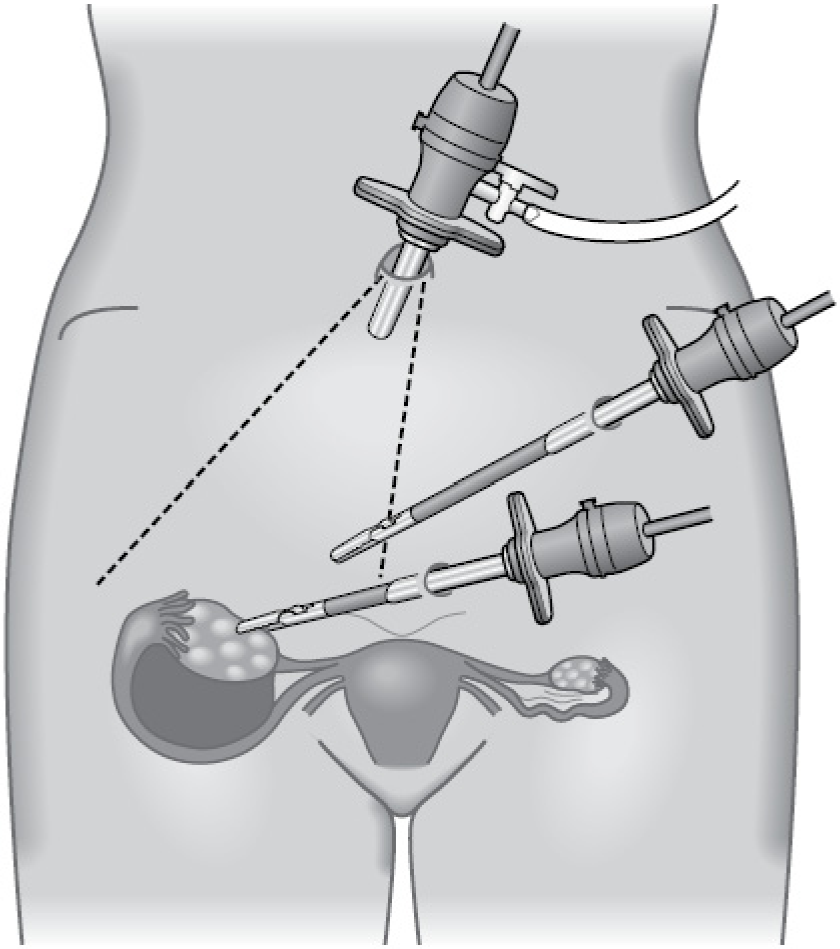
OPERATIVE PROCEDURES FOR NON-NEOPLASTIC OVARIAN CYSTS

Anesthesia

General endotracheal anesthesia with muscle relaxation is required. For both open and laparoscopic procedures, decompression of the bladder with a Foley catheter is recommended to provide optimal access to the pelvis. For laparoscopic procedures, orogastric decompression is also recommended.

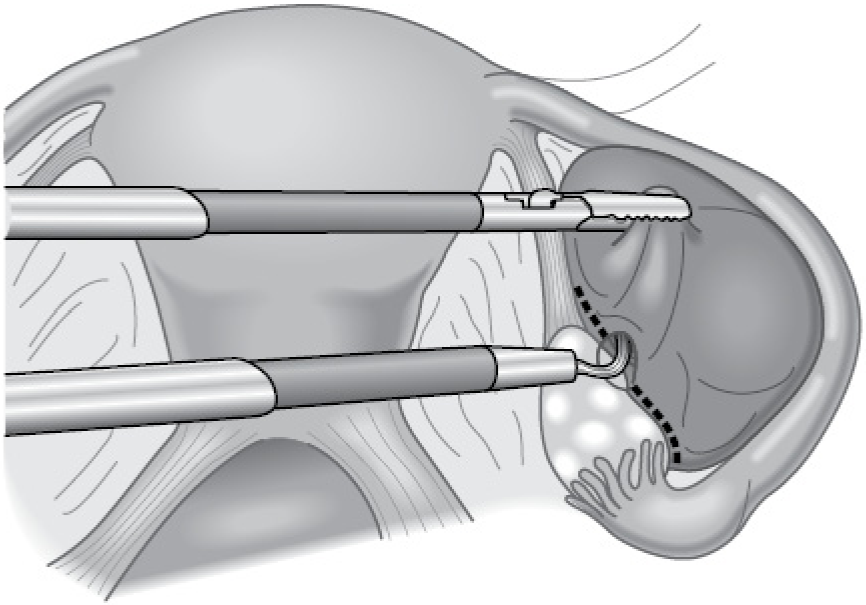
Laparoscopy for non-neoplastic cysts

For laparoscopic cases, a 5 mm umbilical port is placed for insufflation and inspection with the camera. This may be upsized to a 10 mm or 12 mm port if necessary to facilitate utilization of a retrieval bag. Two additional 5 mm trocars are placed for grasping and manipulation (**Figure 81.3**). Placement of the working ports is best dictated by the ability to triangulate the lesion. In general, this involves a suprapubic port and a contralateral lower quadrant port. Transabdominal aspiration under direct vision with a long 18-gauge spinal needle may allow the cyst to be grasped more securely. A portion of the cyst wall is excised with hook cautery to create an adequate fenestration, and then a portion submitted for pathologic evaluation.



81.3

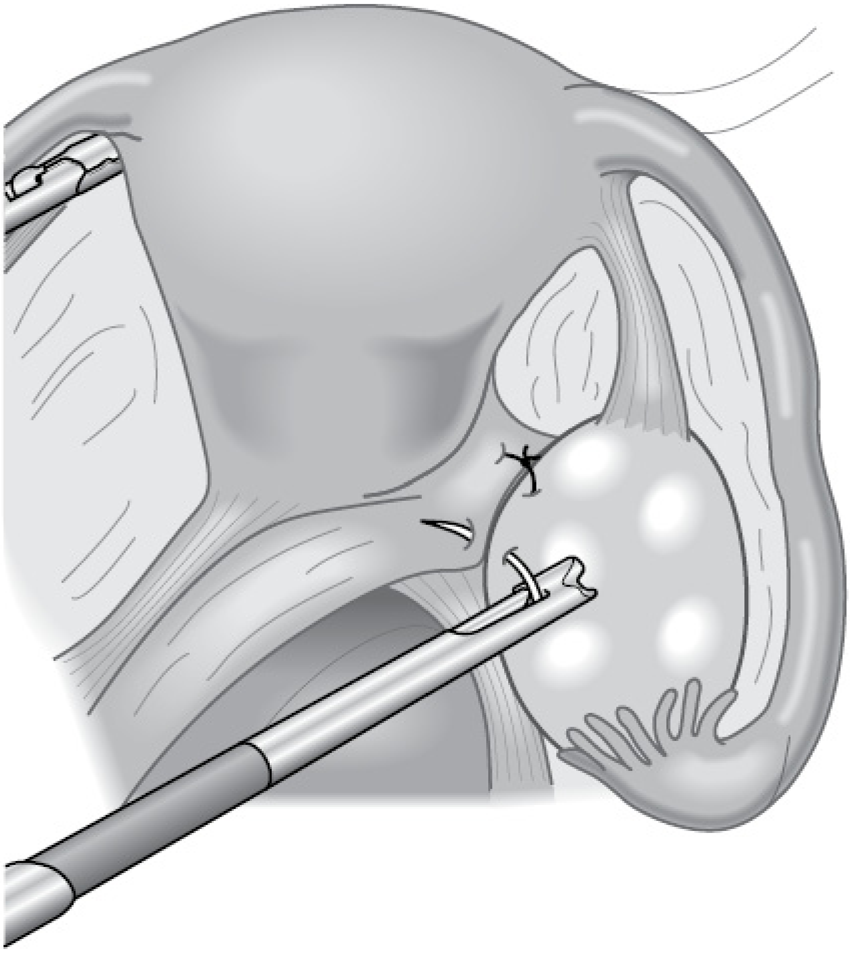
When the cyst is clearly demarcated from the ovarian parenchyma, cyst enucleation is the best option. This can be done with a hook cautery or LigaSure™ (Covidien, Mansfield, MA, USA). The peritoneum is incised circumferentially to demarcate the cyst attachment to the underlying parenchyma (**Figure 81.4**). The cyst is then easily enucleated. Occasionally, laparoscopy for presumed ovarian pathology reveals a paratubal cyst. These are remnants of the paramesonephric duct. They may lie within the leaves of the mesosalpinx or as a pedunculated cyst near the fimbria, and may have evidence of torsion. These lesions may be resected, enucleated, or unroofed.



81.4

Surgical therapy for ovarian torsion

Ovarian torsion may occur with a normal ovary, ovarian cyst, or ovarian tumor. US frequently demonstrates multiple small peripherally located, uniform cysts in the involved ovary. This appearance arises secondary to displaced follicles, due to venous congestion and edema. Color Doppler may reveal absence of arterial flow. A twisted pedicle may also be seen. In cases involving simple cysts, management of the cyst with ovarian preservation is the goal of therapy. Since it is not possible to differentiate benign from malignant lesions in the acute setting of torsion, the principles of ovarian preservation are still valid. If a neoplasm is present, management should be as directed in the section on ovarian neoplasm below. In the absence of evidence of neoplasm, it is preferable to untwist the ovary, and consider oophoropexy to allow resolution of inflammation and edema. This can be accomplished by detorsion, then fixation of the ovary with absorbable suture through the tunica of the ovarian pole to the adjacent utero-ovarian ligament, or to the psoas tendon (**Figure 81.5**). Care must be exercised to identify the ureter prior to suture fixation. Patients should be followed by close postoperative surveillance of the ovary with US in 6–8 weeks.



81.5

OVARIAN NEOPLASMS

Ovarian neoplasms often have a cystic component. The overall risk of malignancy for ovarian neoplasms in children and adolescents is approximately 20%. Pelvic US, although useful in confirming the presence of neoplasm, is not helpful in determining pathologic nature. There are no gross characteristics that clearly distinguish benign from malignant neoplasms and therefore it is best to approach all ovarian neoplasms as if malignancy may be present.

US findings suspicious for neoplasm should also be evaluated by computed tomography (CT) scan to assess for adenopathy, hepatic involvement, and presence of tumor elsewhere within the peritoneal cavity. Tumor markers alpha-fetoprotein (AFP), beta-human chorionic gonadotropin (β-HCG), and inhibin should be determined preoperatively. AFP is elevated in tumors that contain the malignant germ-cell element endodermal sinus tumor and β-HCG is elevated in tumors that contain the malignant element choriocarcinoma. Inhibin is secreted from the ovarian follicle, and may be elevated with granulosa cell tumors and mucinous subtype of epithelial carcinoma.

Although elevated tumor markers are associated with malignancy, they may be normal in up to 45% of malignant pediatric ovarian masses. Epithelial-based tumors may have elevation of CA125.

OPERATION FOR OVARIAN NEOPLASM

At present, there is insufficient evidence to support routine laparoscopic management of ovarian tumors with features to suggest malignancy. There is a recent trend to laparoscopic resection of early (stage 1) ovarian tumors. However, the available data, primarily from the adult literature, suggest the incidence of tumor rupture is unacceptably higher in the laparoscopy (65–100%) group versus the laparotomy (5–10%) group, for both oophorectomy and for enucleation. Intraoperative tumor rupture converts a stage 1 to a stage 2 tumor, with accompanying changes to the treatment protocols. For tumors undergoing laparoscopic resection, it is imperative that the tumor be resected intact, and a retrieval technique (Endocatch, Covidien) be utilized that avoids intraperitoneal spillage. Complete staging (see below) is mandatory. The risk of malignancy in pediatric ovarian neoplasms, the ability to complete a full staging procedure, and the size of the tumor make an open approach preferable in most cases.

Laparotomy for ovarian neoplasms

The choice of incision depends on the size of the lesion and the surgeon's preference. In general, a lesion with a diameter less than the span of the iliac crests may be successfully approached by a transverse incision in the lowest skin crease, or a muscle-sparing Pfannenstiel incision. Lesions exceeding this size will best be approached by a midline incision. The procedure should then follow a systematic assessment, as follows.

PERITONEAL CYTOLOGY

Immediately upon entering the peritoneal cavity, any fluid in the pelvis should be aspirated for ascitic cytology. If no fluid is present, pelvic washing should be done with normal saline.

INSPECTION OF THE OVARY

The involved ovary should be inspected for resectability.

COMPLETE ABDOMINOPELVIC EVALUATION

The remaining staging maneuvers should then be completed for all neoplastic lesions. These include:

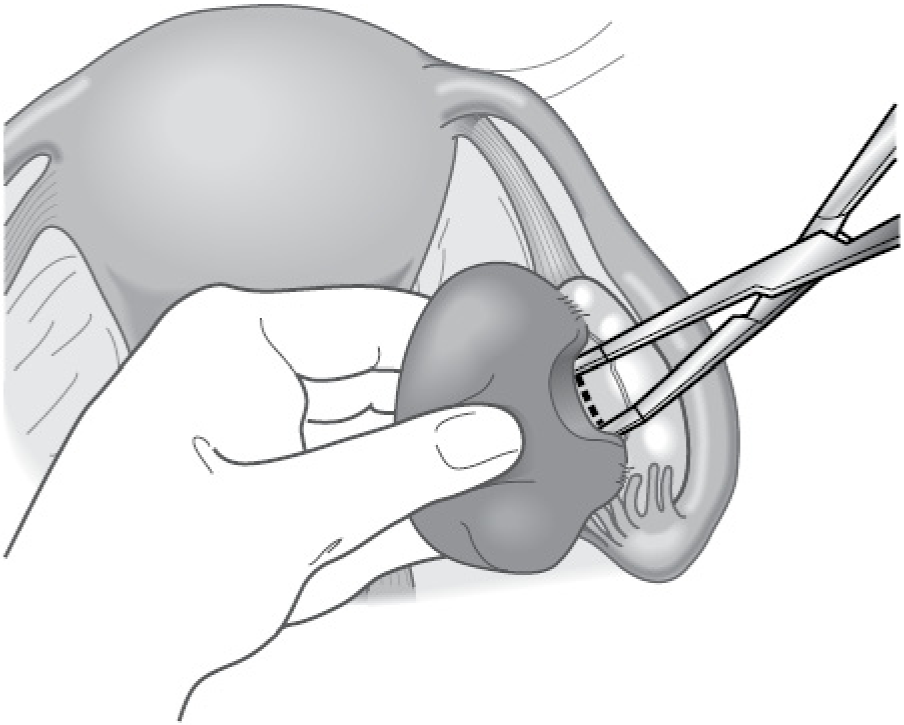
- Inspection and palpation of the omentum, with removal of any abnormal areas
- Inspection and palpation of the retroperitoneal lymph nodes from the iliac chains up along the aorta and vena cava, at least as far as the renal pedicle; any enlarged or firm nodes should be biopsied
- Inspection of the pelvic and visceral peritoneal surfaces with biopsy of any abnormal areas
- Inspection and palpation of the contralateral ovary with enucleation of any abnormal masses; it is not necessary or recommended to bisect a contralateral ovary that appears normal

- In the context of epithelial (mucinous) tumors, an appendectomy should be considered at the time of surgery

Management of the ovarian mass

ENUCLEATION OF OVARIAN MASS

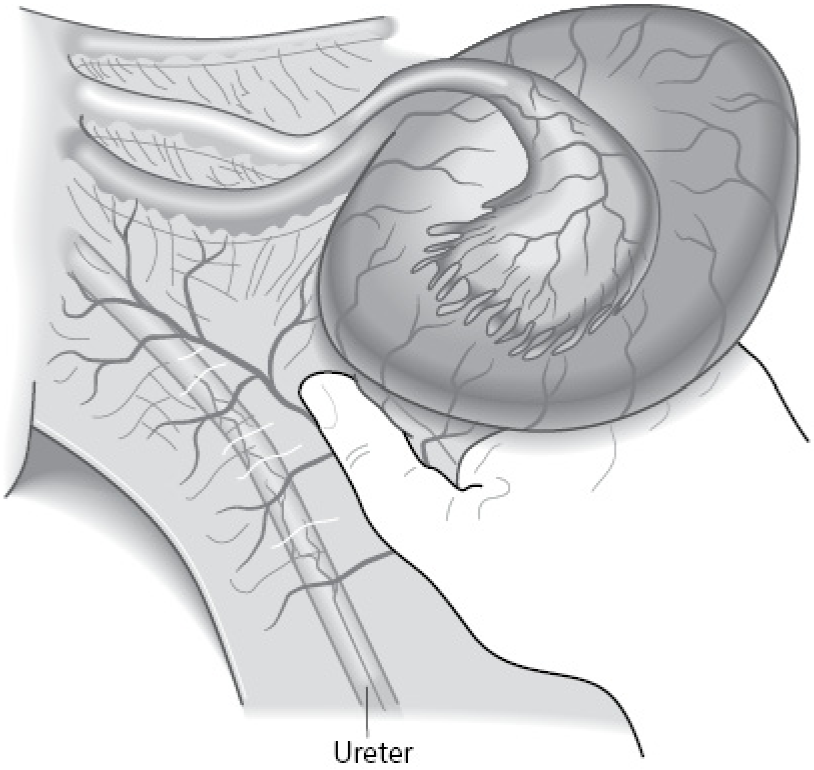
Enucleation of ovarian neoplasms may be considered for smaller masses with normal markers and a clear dissection plane from normal ovarian tissue, or in patients with bilateral suspected neoplasms with visible residual normal arenchyma. If no visible dissection plane is seen on the capsule but a focal mass lesion can be felt, the ovarian cortex is incised to expose the border between the parenchyma and the mass. Dissection should remain outside the capsule of the neoplasm with care not to rupture it (**Figure 81.6**). The ovarian capsule edges may be reapproximated with absorbable suture to minimize raw surface area.



81.6

If the tumor markers are elevated or there is no demarcation between normal ovary and neoplasm, oophorectomy should be undertaken. In most cases, the Fallopian tube is not involved and may be spared.

With large tumors, the leaves of the mesovarium are widely splayed and the Fallopian tube may be draped over the mass. The peritoneum of the mesovarium is incised in an avascular plane between the tube and ovary with cautery dissection. This maneuver will expose the vascular pedicle of the ovary. If the tumor is large, the venous plexus is often engorged and the pedicle may need to be taken in stepwise fashion with pairs of clamps, division of the tissue, and ligation. The assistant should maintain manual compression of the pedicle as the dissection proceeds (**Figure 81.7**). If the tube is adherent, it may be taken in continuity with the ovary. Caution should be taken to clearly identify the ureter before proceeding with division of the mesosalpinx.



81.7

BIOPSY

If the lesion is invading the uterus or adjacent pelvic structures, it should not be resected at the primary operation. A biopsy for tissue diagnosis should be done with a plan for later resection after chemotherapy. Chemotherapy is highly successful in producing shrinkage of tumors to allow later resection without sacrifice of uterus or other pelvic structures.

POSTOPERATIVE CARE

Regardless of approach chosen, the Foley catheter can generally be removed immediately after the procedure for small- to moderate-sized lesions and rapid advancement of diet is tolerated. For large lesions requiring a generous incision, it may be more comfortable for the patient to leave the Foley catheter in place for 24 hours. If a large tumor requiring a generous incision is present, supplemental epidural catheter for postoperative pain management should be considered.

OUTCOME

Fenestration for non-neoplastic cysts has excellent outcome with minimal long-term consequences. Malignant germ-cell tumors are treated with platinum-based multiagent chemotherapy. Survival with current regimens is greater than 90% at 5 years.

All neoplastic lesions, whether benign or malignant, have some risk of contralateral recurrence. This is particularly true for ovarian teratoma, where more than 20% of children with mature ovarian teratoma will develop metachronous tumors to the contralateral ovary. Therefore, yearly US follow-up is needed for these patients, to allow detection of new lesions at an early stage so that an ovary-sparing procedure may be undertaken with greater chance of success in preserving endocrine function and fertility. Tumors with positive markers should be followed with postoperative serum markers to help detect recurrence. Epithelial malignant neoplasms should adhere to adult treatment and follow-up protocols due to the rare nature of these tumors.

ACKNOWLEDGMENTS

Figure 81.1 was obtained from Riley Childrens's Hospital, Indianapolis, IN, USA. Courtesy of Dr. Boaz Karmazyn.

FURTHER READING

Billmire D, Vinocur C, Rescorla F, et al. Outcome and staging evaluation in malignant germ cell tumors of the ovary in children and adolescents: An intergroup study. *J Pediatr Surg* 2004; 39: 424–9.

Brandt ML, Helmrath MA. Ovarian cysts in infants and children. *Semin Pediatr Surg* 2005; 14: 78–85.

Chang HC, Bhatt S, Dogra VS. Pearls and pitfalls in diagnosis of ovarian torsion. *Radiographics* 2008; 28: 1355–68.

Morowitz M, Huff S, von Allmen D. Epithelial ovarian tumors in children: A retrospective analysis. *J Pediatr Surg* 2003; 38: 331–5.

Nabhan ZM, West KW, Eugster EA. Oophorectomy in McCune–Albright syndrome: A case of mistaken identity. *J Pediatr Surg* 2007; 42: 1578–83.

Song T, Choi CH, Lee Y-Y, et al. Pediatric borderline ovarian tumors: A retrospective analysis. *J Pediatr Surg* 2010; 45: 1955–9.

Taskinen S, Urtane A, Fagerholm R, et al. Metachronous benign ovarian tumors are not uncommon in children. *J Pediatr Surg* 2014; 49(4): 543–5.

PART VIII

Trauma

82

Burns and burn surgery

HEINZ RODE AND ROUX MARTINEZ

HISTORY

Although members of the Indian Tilemaker caste have been credited with attempts at skin grafting between 2000 and 3000 years ago, it was not until the middle of the 19th century that serious attempts were made to transpose skin from one part of the body to another. George Pollock of St George's Hospital in London performed the first "pinch" skin grafts for a burn in 1870, and Karl Thiersch used more extensive pieces of split skin for grafting in 1874. Free full-thickness grafts were first used to repair eyelid defects by the Hungarian-born but Scots-trained John R. Wolfe in 1875.

Lustgarten described a technique of early excision and grafting of small burns in 1891. Brown of St Louis established the practical aspects of biological skin dressings as life-saving procedures in extensively burned patients. However, treatment remained rudimentary until the middle of the 20th century when early tangential excision and immediate autologous skin grafting began to be introduced by Zora Janžekovič (1918–2005) working in the small Slovenian town of Maribor, then in Yugoslavia.

PATHOPHYSIOLOGY

Burn injuries are extremely complex and elicit pathophysiologic and metabolic responses involving all major organ systems. Although there are different injury patterns, the basic pathophysiology involves local and systemic responses to excessive energy transfer of liquid, fire, contact heat, radiation, and chemicals.

- *Local response:* Jackson in 1947 described the classical three zone responses to burn injury. These zones are three-dimensional and consist of:
    - The *zone of irreversible coagulation* is at the point of maximum energy transfer to the skin and underlying tissues.
    - The *zone of stasis* surrounds this, characterized by decreased tissue perfusion and is potentially salvageable.
    - The *zone of hyperemia,* the most peripheral zone, is potentially viable, provided resuscitation is adequate and infection prevented.
- *Systemic response:* In patients with moderate to major burns, inflammatory mediators are released into the systemic circulation, causing a systemic inflammatory response characterized by cardiovascular, respiratory, metabolic, and immunological changes. These changes become more pronounced the larger the burn, resulting in hypotension and end-organ hypoperfusion, increased susceptibility to infection, energy depletion, hypermetabolic response, and alterations in pulmonary function similar to those seen with other forms of traumatic injury.

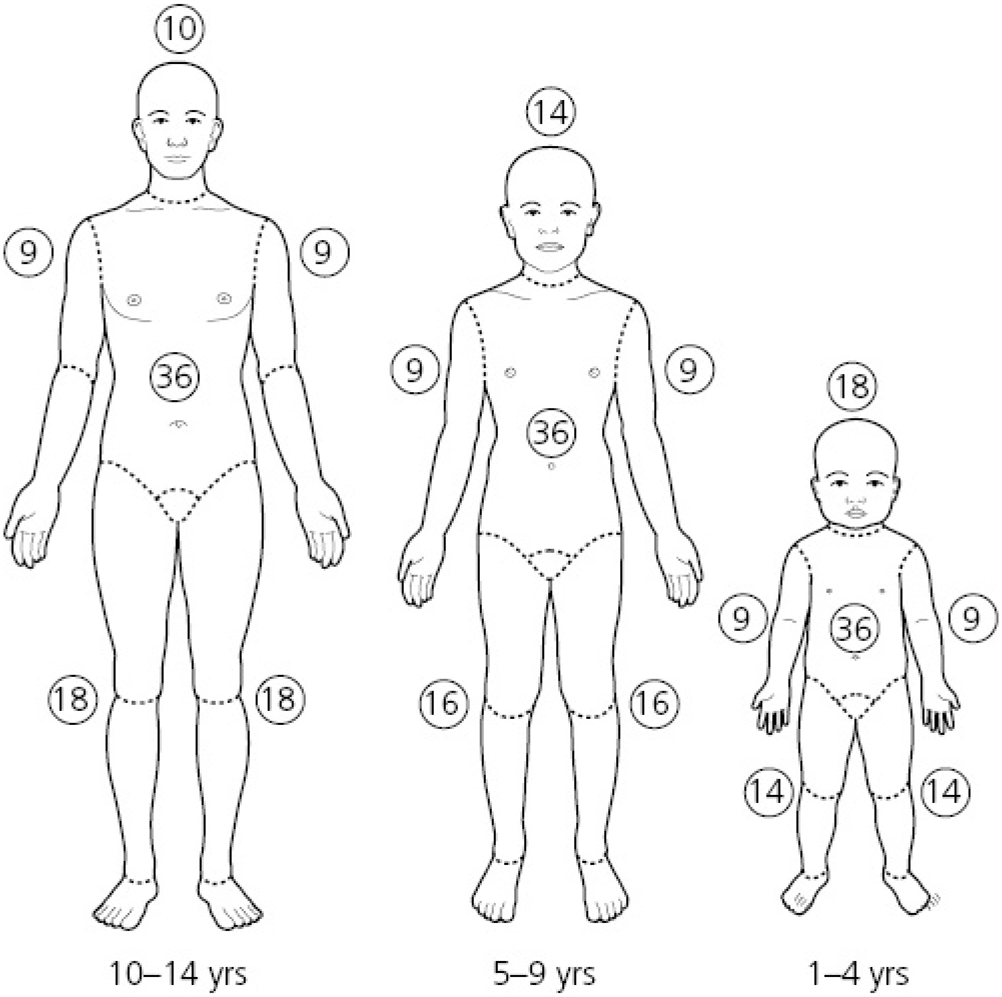
BURN SEVERITY

Five key factors determine appropriate therapy:

- Extent – expressed as a fraction of the total body surface area
- Depth of the burn
- Anatomical area of involvement
- Concomitant diseases or injury
- Age and physiologic status of the patient

Estimating the extent of the burn

There are important proportionate differences in body surface related to the age of a child. For those aged 10 years and above, the "rule of nines" popularized by Wallace should be used (**Figure 82.1**). For those <10 years, more specific "pediatric" charts (e.g. Lund and Browder) should be consulted.



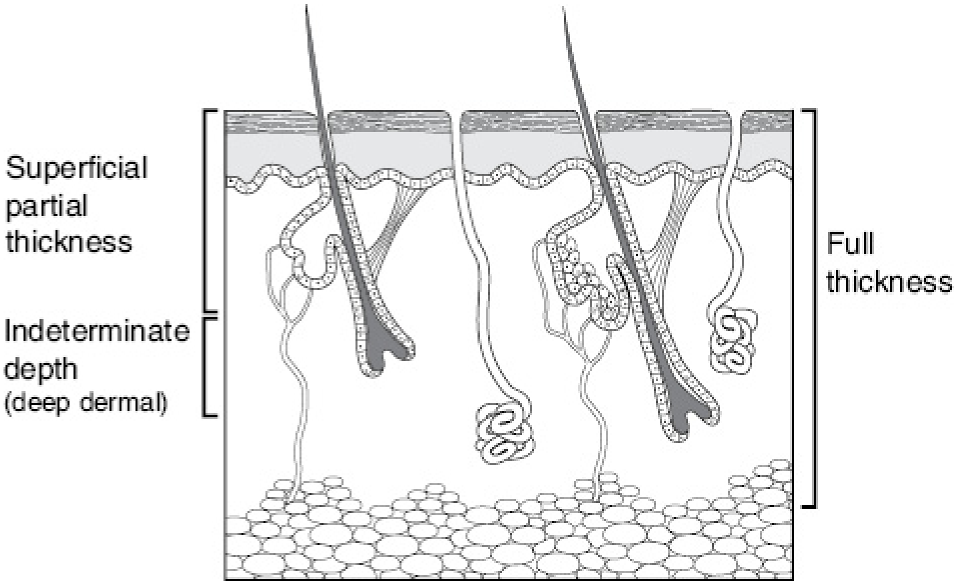
82.1

As the palmar surface of the open hand amounts to ~1% of total body surface area (TBSA), this can be used to estimate the size of small or scattered burns in children.

Modern smartphone apps are available as an accurate mobile device to determine the TBSA as well as a visual recording of the injury (e.g. the Rapid Burn Assessor).

Estimating the depth of the burn

There is no objective clinical method available to determine the depth of a burn, unfortunately. Most burns are a combination of superficial and deep burns (**Figure 82.2**), and the best assessment should be made 3–4 days after the injury, when wound evolution has occurred (**Table 82.1**).


Superficial partial thickness
Indeterminate depth (deep dermal)
Full thickness

82.2 Levels of burn injury.

Table 82.1 Assessment of burn depth

Burn depth	Physical appearance
Superficial partial thickness	
Destruction of only superficial epidermis of the skin Spontaneous epithelialization within 3 weeks Excision is contraindicated and the wounds rarely cause functional or cosmetic defects or hypertrophic scars	Erythematous, moist, and painful with blisters Hypersensitive to touch and blanches readily
Deep partial thickness (indeterminate depth)	
Destruction of epidermis *plus* varying amounts of dermis These wounds are difficult to assess during the first 3 days after injury due to the ongoing evolution within the burn wound The healing time for these wounds may be variable and may require several weeks leading to severe hypertrophic scarring	Reticulated red, white dry surface and may blister Capillary circulation may be sluggish or absent when pressure is applied
Full thickness	
Irreversible destruction of all elements of the skin ± extension into deeper tissues and structures Prolonged healing (>3 weeks) with unsatisfactory functional and cosmetic results Aim for early eschar excision and immediate grafting	May mimic the appearance of an indeterminate burn and are usually mottled, white, red, or charred and dry in appearance Insensitive to pain and leathery to palpation with no capillary return Blisters are unusual and, if present, are thin-walled and do not enlarge Clotted superficial vessels may be visible Appearance of the burn remains static with little change over the ensuing days

During the first few days, clinical judgment is based on visual and tactile characteristics of the burn wound. A combination of clinical evaluation, laser Doppler image scanning, and indocyanine green (ICG) video angiography is recommended for burns of undetermined depth. Punch biopsies can also be used, but sampling errors and histological interpretation have shortcomings.

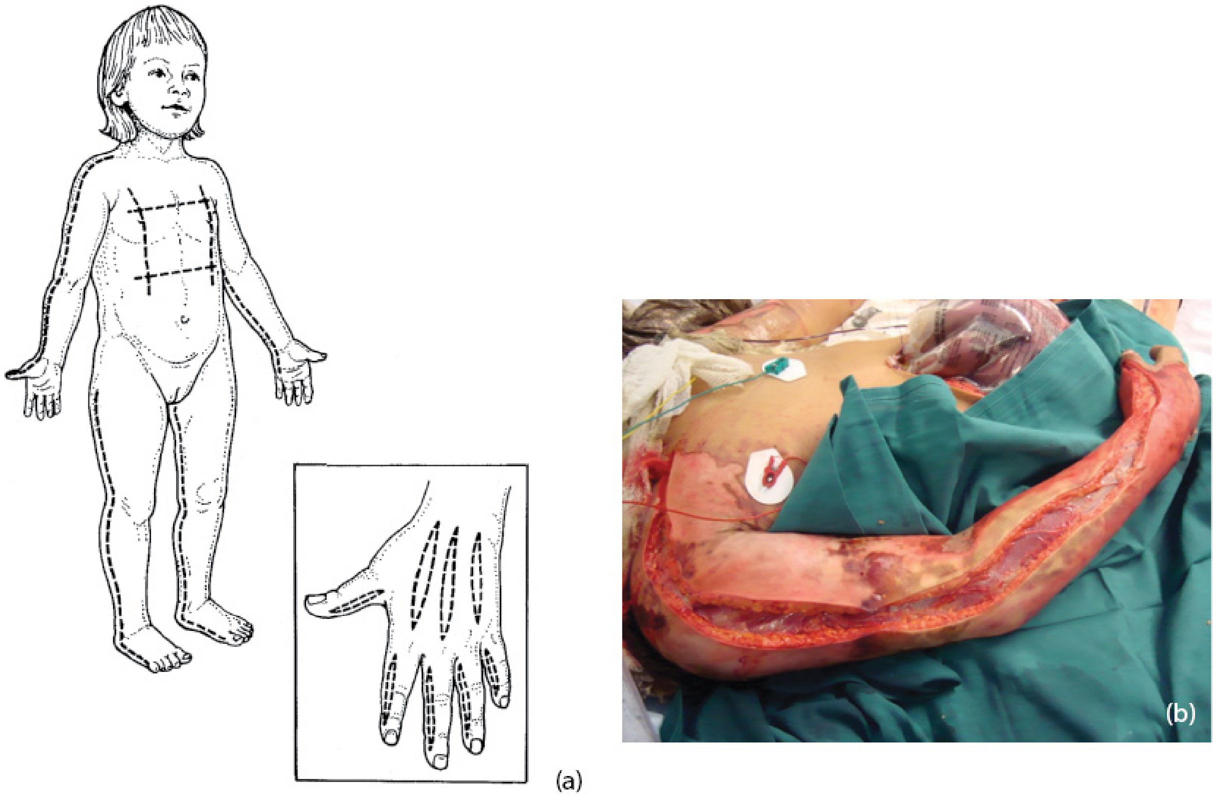
MANAGEMENT

Emergency management

- *On-site:*
  - Ensure self-safety
  - Remove smoldering or hot clothing and ensure airway patency
  - *Primary survey* to identify and treat life-threatening conditions
  - *Secondary head-to-toe survey* and management if required
- *Hydrotherapy:* Immerse small burns (<25% TBSA) in cold or running water (15–18 °C) or cover with cold, wet compresses for at least 30 minutes to relieve pain and discomfort; cold water should be used cautiously for larger burns to prevent hypothermia
- *Chemical burns:* Also require copious irrigation with water (at least 30 minutes); avoid "neutralizing agents"
- *Transport*: Cover burn wounds with a clean plastic sheet
- *Electrical burns:* Evaluate and maintain the circulation and ventilation throughout
- *Inhalation injury:* Give 100% oxygen by face mask; progressive airway obstruction may occur over the first hours after the burn; it is safest to intubate early, before extensive head or laryngeal swelling occurs and airway obstruction becomes imminent
- Withhold oral fluids initially
- Stabilize patient before transportation is undertaken

Decompression escharotomy

This is an emergency procedure done at the time of the first assessment to avoid irreversible ischemic and hypoxic damage to tissue. The decision is a clinical one but should be considered in circumferential deep burns that are impeding circulation to more distal parts – typically arms, hands, legs, and even the chest (**Figure 82.3a**). It is ideally performed in operating room under general anesthetic with diathermy or scalpel (**Figure 82.3b**), but it can also be done at the bedside. It is important not to damage nerves and tendons during the procedure, which must be extended from above to below the constricting eschar and be skin deep into bulging fat.



82.3a,b (a) Typical areas of the body where deep burns might need decompression escharotomy. (b) Circumferential full-thickness burn of upper limb requiring decompression escharotomy.

It is prudent to monitor the return of distal circulation and improvements in chest excursions. With “fluid creep,” a “compartment syndrome” may develop in the extremities, necessitating further decompression.

DEFINITIVE TREATMENT

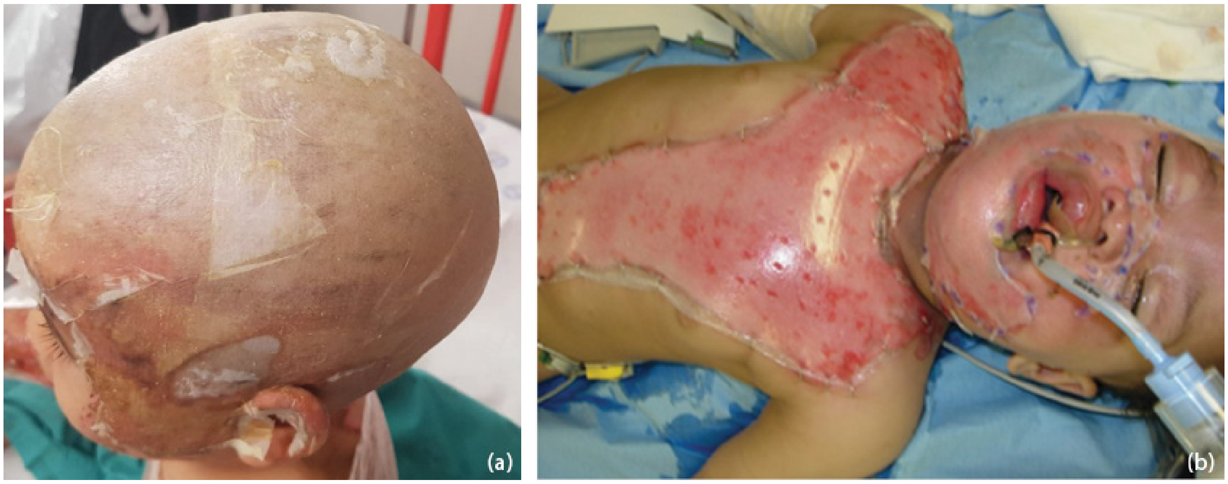
A distinction should be made whether the wound should be treated conservatively or whether surgical excision is required. Management is therefore principally divided into those burn wounds that will re-epithelialize spontaneously by primary intention and those that require excision and grafting.

Superficial partial-thickness burns

Epidermal and superficial partial-thickness burns are treated non-operatively and will heal provided resuscitation was adequate and burn wound infection was either prevented or treated effectively. They should heal within 14–28 days. Skin grafting will not improve functional outcome, time to healing, or esthetic result.

Clean the wound and remove all loose devitalized or contaminated tissue including blisters and any topical agents and exudate. Wash with a bland soap followed by the topical application for 20–30 minutes of 0.006% sodium hypochlorite (NaOCl) solution or equivalent for 20 minutes. Dry the wound and then apply either a short- or long-acting topical antiseptic with an occlusive dressing.

Alternatively, the wound can be covered with synthetic or biosynthetic membranes (e.g. Biobrane®, Nanotrix®, KeraGENtrix®, or Suprathel®) (**Figure 82.4a and b**). The latter have many advantages over the traditional antiseptics. They require a viable bed, are applied once, and firmly adhere to the wounds. They absorb exudate and are semipermeable barriers to evaporative losses and protect the wounds from desiccation and contamination, are permeable to topical antiseptics, stimulate re-epithelialization, and improve patient comfort.



82.4a,b Management of deep partial- and full-thickness burns: applying synthetic skin substitutes.

The membrane can be applied with or without anesthesia. The burn wound must be prepared by removing all loose and necrotic tissue and be vascularized, and hemostasis must be complete. The preferred biosynthetic wound cover is then applied to the burned area under slight tension and secured in place with strips of adhesive tape or tissue glue. It is then covered with an antiseptic (Xeroform®) followed by an outer dressing. The outer dressing is removed after 24 hours and any fluid accumulation removed. As the wound heals by primary intention, the synthetic material gradually peels off within 2 weeks as re-epithelialization progresses. None of these membranes has any inherent antimicrobial properties and wound infection remains a risk.

Deep-dermal and full-thickness burns

These burns represent a unique challenge due to the nature of the operation and the concomitant pathophysiological responses to injury. The process of excision, grafting, and burn wound coverage must follow a predetermined progression.

PREOPERATIVE PREPARATION

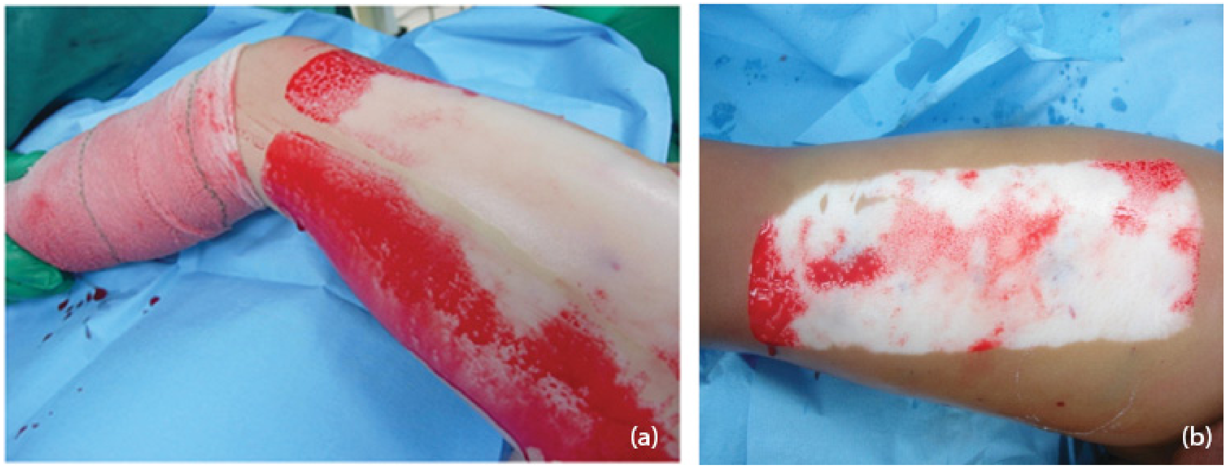
Operating room and anesthetic requirements

- The patient must be hemo- and thermodynamically stable and any physiologic abnormalities must have been corrected.
- Premedication must include sedatives and anxiolytic agents to alleviate anxiety and fear. It is withheld from infants and those on IV morphine infusion and during emergency procedures.
- Assess lung function and any effect of the burn or inhalation on the upper airways before the induction of anesthesia.
- Avoid intraoperative hypothermia, keeping the patient's core temperature >37 °C. Maintain the ambient operating room temperature at 28–32 °C and warm all IV fluids.

- Intraoperative antibiotics are not administered unless the patient has a compromising systemic disease or invasive burn wound infection, or when a major burn excision was performed.
- Vascular access is essential and an arterial line is required for large burn excisions.
- Blood loss can be rapid and substantial from both the excised and donor areas.
  - Replace with packed cells, but with massive hemorrhage, fresh frozen plasma or other components may be required.
  - Blood loss can be reduced by pre-excisional subeschar injection (clysis) (**Figure 82.5a and b**) of local anesthetic and adrenaline to both the area for excision and the selected donor site. The formulation for clysis is given in **Table 82.2**. Other methods include local pressure, diathermy coagulation, suture ligation, calcium-enriched alginate dressings, pneumatic tourniquet, topical application of 1 : 10 000 or 1 : 30 000 adrenaline solution and the temporary application of the procured autograft. Topical adrenaline should not be used in patients with cardiac pathology or arrhythmias.

Table 82.2 **Clysis formulation**

Bupivacaine 0.5% 10 mL (50 mg)
Adrenaline (Epinephrine) 1 mg/1 mL (1:1000)
Combine to a total of 11 mL
Take 4.4 mL of this mixture and add to 200 mL Normal Saline or Ringer's Lactate
Use up to a maximum of 20 mL/kg body mass



82.5a,b Pre-excisional subeschar injection (clysis).

Preparing the burn wound for surgery

It is necessary to carry out a preliminary head-to-toe wash, including the donor sites and the burn wound, with 1% chlorhexidine soap solution followed by the topical application of 0.006% sodium hypochlorite solution for 20 minutes.

PRACTICAL APPROACH TO SURGICAL EXCISION

- In general terms, excision is carried out as soon after injury as possible, i.e. as soon as the cardiovascular system has been stabilized and resuscitation completed. This time may vary from a few hours to several days, but a good timing goal is day 3.
- Excision is an elective procedure in a stable patient for deep-dermal and full-thickness burns. Burns of an indeterminate depth are best observed for 10–14 days and, if anticipated that it will not be healed by 3 weeks, should then be excised and grafted.
- The amount to be excised at each procedure depends on the stability of the patient, the burn size, availability of auto- and allografts, the volume of blood loss incurred during the procedure, and the adequacy of anesthesia.

- A decision must be made about the area and extent of the burn to be excised. It is advisable to limit excision to 25–100 cm^2, control hemorrhage, and then to proceed with further excision. A reasonable total operative time is 2–3 hours.
- The sequence of areas to be grafted is variable and depends on individual preferences.

It is preferable to do several smaller procedures, 1–2 days apart, rather than one major excision that can compromise the safety of the patient.

PRINCIPLES ACCORDING TO DEGREE OF BURN AREA

- *Burns <10% TBSA:* Excision and autografting are performed using meshed 1.5 : 1 or 2 : 1 or sheet grafts. Sheet grafts are placed on all vital and visual areas (face, neck, chest, hands) and all small grafted areas.
- *Burns 10–30% TBSA:* Excision and autografting are required.
- *Burns 30–40% TBSA:* Sufficient donor sites are usually available to graft the excised bed, despite the fact that about 30% TBSA is unavailable for donation (face, neck, hands, and feet). The grafts should be meshed at 1.5 : 1 or 2 : 1, or temporary allografts or a synthetic skin substitute applied, until definitive autografting can be performed.
- *Burns >40% TBSA:* There is major limitation of donor sites, therefore alternative methods are required. These include the wide expansion of autografts (1 : 3–1 : 9), repeated skin harvesting from the same area (two or three times at most), the modified “Meek” micrografting technique, the use of temporary biosynthetic skin substitutes, allograft, xenografts, and, in rare circumstances, cultured epithelial grafts.
- The modified “MEEK” technique of multiple skin “postage stamp” grafts is gaining rapid acceptance as the method of choice in major burn centers to achieve primary epithelial/dermal skin cover. These micrografts contain both epidermal and superficial dermal components and can be expanded to a maximum ratio of 9 : 1. The recipient bed must be viable and non-infected and can be prepared by the temporary application of allografts. The technique is not suitable for facial, neck, perineal, or hand grafts.

- In rare circumstances, cultured epithelial autografts can be used as either "spray-on" or "sheet" grafts. The latter method is only used as a rescue method in a massive, life-threatening burn. Complications are numerous, being extremely fragile and less pigmented. Over half will need further grafting, and they have a tendency to contract with a lifelong risk of malignancy.

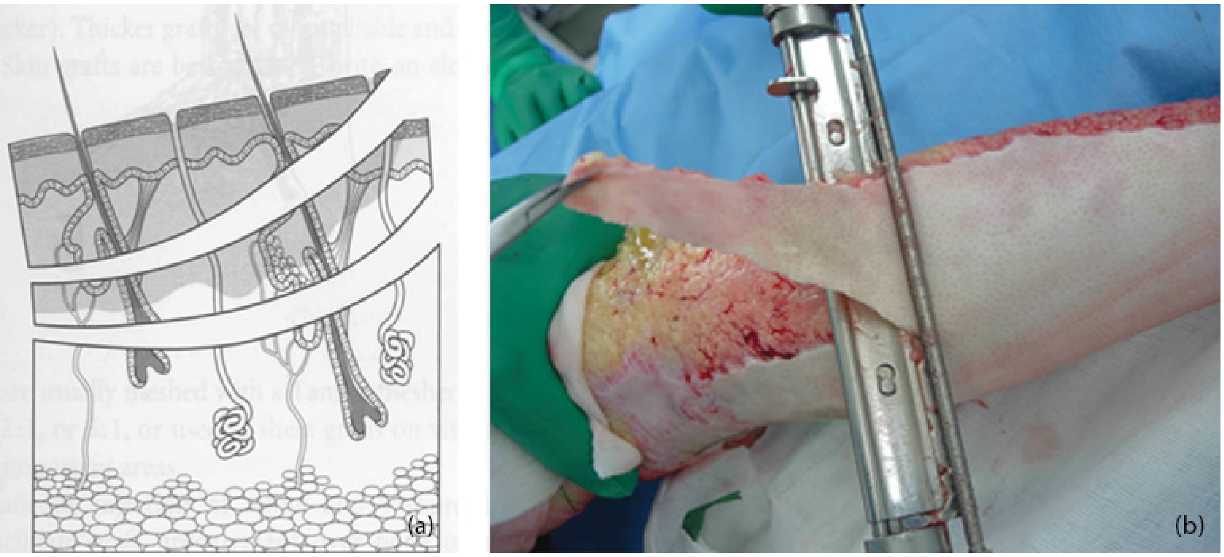
SURGICAL TECHNIQUE

A variety of methods are used to remove the burn eschar, depending on the depth of the wound, the time post-burn and the changes occurring in the eschar over time.

- Tangential, sequential, or layered excision (procedure of choice in children)
- Fascial excision
- Delayed escharectomy
- The use of synthetic and biological skin substitutes
- Dermal substitution

Tangential excision

This entails the sequential excision of thin layers of burn eschar until viable dermal or subcutaneous fat is reached (**Figure 82.6a**). If done correctly, the minimum amount of living tissue is sacrificed and dermal elements are retained, with satisfactory functional and cosmetic results. Tangential excision should be done within the first 1–5 days before hypervascularity and wound infection become established.



82.6a,b Tangential excision.

Excision is best performed by using a hand-held Humby or Watson knife, held at a tangent to the wound surface (**Figure 82.6b**). The depth of each back and forward motion of the knife is controlled by the angle of cutting and the thickness of the guard been used. Excision over uneven surfaces or bony prominences can be aided by a subeschar injection of saline. Smaller areas can be excised with a Goulian knife.

The appropriate level of excision or end point is characterized by a shiny white surface with brisk arteriolar or punctuate bleeding or viable yellow, non-hemorrhagic subcutaneous fat globules with briskly bleeding vessels in all areas. Clotted vessels, dullness, punctate hemorrhages, dark pinkish-brown hemorrhagic fat represent non-viable areas and must be removed as residual necrotic areas will jeopardize graft take. Tissue staining with hemoglobin from lysed red cells indicates inadequate excision. Dissection may become difficult once the level of excision has gone beyond the dermis. The correct level of excision is crucial. If there is doubt about the viability of the level of excision, it is preferable to re-excise a day or two later.

Once excised to the appropriate level, and hemostasis is secured, an immediate split-thickness autograft should be performed or a skin substitute used as a temporary biological wound cover. In burns <40%

TBSA excision and grafting can be done in one procedure; in unstable patients, further excisions may be delayed 24–48 hours to complete the procedure.

The topical application of adrenaline also changes the clinical appearance of the excised wound and the surgeon must be certain of the viability of the recipient bed before embarking on further excisions.

Fascial excision

This procedure is generally reserved for very large, life-threatening (>60% TBSA) or deep full-thickness burns during the first few days following the burn. By incising at the periphery of the eschar vertically downward to the level of the viable deep fascia, a flap of eschar is raised and the dissection extended until all the dead tissue has been excised.

The excision is performed using a combination of sharp dissection, traction, and hemorrhage control or cautery. It is preferable to leave a thin layer of fat over the subcutaneous bony prominences and tendon sheaths.

Fascial excision usually assures a viable bed for skin grafting with only moderate blood loss, especially if under tourniquet control. The skin edges are sutured to the excised bed to limit movement and control bleeding, and the excised area is covered with expanded split-thickness skin grafts. If the ratio exceeds 2 : 1, the autograft should be covered with a 1.5 : 1 meshed allograft, or skin substitutes, and “take” will be improved by the use of negative-pressure wound therapy.

The major disadvantage of this method is that it causes damage to lymphatics and cutaneous nerves, loss of subcutaneous fat, long-term cosmetic deformity, and distal edema.

Delayed escharectomy

This is the excision of disintegrating, infected, old adherent eschar not previously removed and the removal of overgranulated areas from old neglected unhealed burn wounds. The tissue is not suitable for skin grafting and usually heavily contaminated with bacteria.

Exuberant tissue may be removed surgically, with a hydro dissector device (e.g. the Versajet™ hydrotherapy device) or ultrasonic dissectors (particularly useful to remove tissue around the ears, face, eyes, hands, and genitalia). Bleeding is usually brisk, but with pressure stops readily. Nowadays, enzymatic debriding agents are seldom used.

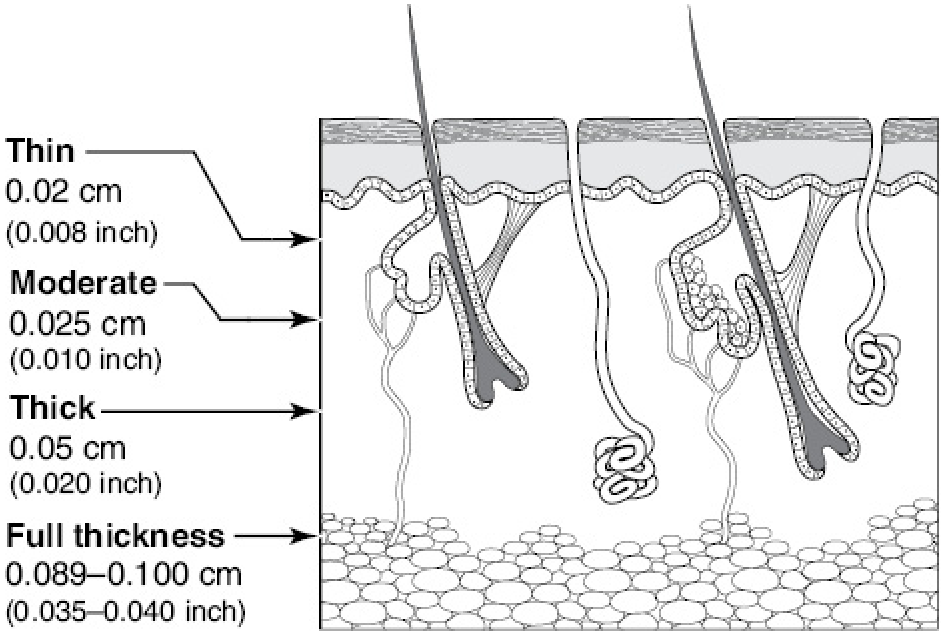
The prepared surface is ready for split skin grafting when there is a shiny, slightly granular pinkish/red uniform bed of granulation tissue with no residual debris or evidence of infection. If still doubtful, a punch biopsy can be performed for histological staining for aerobic, fungal, and viral and biofilm determination.

Harvesting of donor skin

Donor skin may be harvested from any unburned area, although the face, hands, perineum, and cervical areas are unsuitable.

In patients with Fitzpatrick 3–6 complexion (based on proneness to sunburn and ease of tanning), the scalp is contraindicated as a preferred donor site. The preferred sites are the flat surfaces of the legs, buttocks, and back.

Donor skin thickness should be between 0.02 cm and 0.025 cm and should contain both epidermis and papillary dermis (**Figure 82.7**).


Thin
0.02 cm
(0.008 inch)
Moderate
0.025 cm
(0.010 inch)
Thick
0.05 cm
(0.020 inch)
Full thickness
0.089–0.100 cm
(0.035–0.040 inch)

82.7

Harvesting is best performed with an electric dermatome to ensure consistent skin depth, with the skin under tension or with a Watson knife. The subcutaneous injection of saline will aid graft procurement over bony or uneven prominences.

Grafts are usually meshed with a Tanner mesher or blade at a ratio of 1.5 : 1, 2 : 1, or 3 : 1, or used as sheet grafts on more important cosmetic and functional areas such as the face and hands and over joints.

If greater expansion is needed, the *Alexander sandwich technique* may be employed, using temporary skin substitutes (allografts or a synthetic skin substitute) over the widely expanded autograft to prevent desiccation of the interstitial space and achieve primary wound closure.

Although donor sites can be re-cropped every 2–3 weeks, there is a limited number of times as the dermis becomes progressively thinner with each procurement.

Skin graft application

The harvested skin is grafted with the "shiny" or cut side facing the prepared bed. The graft edges should be approximated or slightly overlapping and then secured with surgical staples, fibrin glue, adhesive dressings (Steri-strips, Hypafix®), synthetic tissue glue, or absorbable sutures. If the skin expansion is larger than 1 : 2, it should be covered with allograft or a biological dressing to prevent desiccation of the interstitial spaces, thereby expediting re-epithelialization. A layer of non-adherent fine mesh gauze, impregnated with an antiseptic (Xeroform®), and an outside compression absorbent layer (Kerlix□) is then directly applied to the grafted site.

The grafted area is then splinted in a functional position where applicable. Care must be taken not to wrap the dressings too tightly. Occlusive dressings are required in children to prevent mechanical dislodgement and infection of the grafts. Immobilization of the grafted area is important.

Graft adherence initially is due to fibrin bonds, followed during the next 48 hours by plasmatic imbibition and eventually, after 3–4 days, by vascularization if grafted onto dermis and 5–7 days if grafted on viable fat. The next day all the outer dressings are removed down to the layer of fine gauze to remove any soaked dressing and blood clots and realign displaced grafts. The antiseptic dressing is then replaced and left *in situ* for a few days or changed if it has shifted or there is strike-through exudate or suspected infection.

Securing staples should be removed after 5–7 days and the recipient and donor sites protected with further non-adherent dressings until healed.

Donor site dressing

The split-thickness skin graft donor site is similar to a superficial partial-thickness burn and re-epithelialization can be expected within 2 weeks, depending on the depth of harvest and donor site infection. A number of dressings can be used for the donor sites. The basic dressing should be porous to allow wound exudate and initial postoperative bleeding to exit from the wound surface. The dressings are applied directly on the wound with a few centimeters' overlap. A suitable absorbent outer dressing is then applied.

The primary dressing stays in place throughout the 10–14 day healing process and gradually separates from the donor site as re-epithelialization occurs. The most commonly applied dressings are Xeroform (fine-mesh gauze impregnated with the antiseptic bismuth tribromophenate), DuoDERM® (hydrocolloid), Kaltostat® (calcium alginate), Aquacel® (hydrofiber), Opsite (polyurethane film), Omiderm (synthetic hydrophilized polyurethane wound cover), Mepilex® (silicone-faced polyurethane foam), and Biobrane®.

Graft failure

Graft survival depends on many factors until revascularization occurs at 4–5 days. These include residual necrotic tissue, infected recipient bed, subgraft hematoma or fluid accumulation, mechanical shearing forces, thick skin grafts, and post-graft bacterial infection. The care of the post-transplant wound is therefore of the utmost importance to achieve primary success.

SKIN SUBSTITUTES

Skin substitutes may be used as biological dressings to create a temporary physiologic environment. Some indications for their use are:

- As a primary dressing on clean superficial wounds until epithelialization has been completed
- To provide temporary physiological closure of deep-dermal and full-thickness burns after excision while awaiting autografting
- As sandwich grafts over widely expanded meshed autografts
- As a "test graft" in questionable wound beds
- As dressing on donor sites to facilitate epithelialization and pain control

The most appropriate and gold-standard skin substitute is a human allograft used either fresh, fresh-refrigerated, or glycerol- or cryo-preserved. They achieve an environmental "barrier or seal" of the wound at the graft wound interface, become vascularized, and promote new angiogenesis and maturation of the underlying tissue.

Skin allografts will undergo rejection in 7–14 days and should be replaced with pool allografts or the wound should be autografted when they are available.

If allografts are left for more than 14 days, removal can only be achieved by excision. Other biological skin substitutes at best function only as temporary skin cover and include Integra®, Alloderm®, Biobrane, porcine xenograft, human amniotic membrane, TranCyte®, Oasis Wound Matrix®, and Epicel®.

DERMAL SUBSTITUTES

Dermal substitutes function primarily as biodegradable, 3D scaffolds for dermal regeneration in full-thickness or deep-dermal burns. They provide an adequate environment for the ingrowths of blood vessels and fibroblast over a 3-week period. These dermal regeneration template substitutes can either be grafted immediately with split-thickness skin following their application (e.g. Matriderm®, a structurally intact matrix of bovine type I collagen with elastin) or grafted after 3 weeks with a very thin autograft onto the neodermis (e.g. Integra, which is a bilaminar template with bovine collagen, glycosaminoglycan, and a thin outer layer of silicone). These grafted areas must be inspected daily for fluid accumulation and infection.

Special areas

VOLAR SURFACES

The volar surface of the hand and sole of the foot consists of thick specialized glabrous skin. It is preferable to treat burns in these areas conservatively, awaiting re-epithelialization. Should skin grafts eventually be required, only regional glabrous skin should be used. The thenar and hypothenar areas of the hand and the entire sole and lateral aspects of the foot can be used as donor sites. The ideal graft will be non-translucent and pearly grey. Care is taken not to encroach onto the non-glabrous dorsum surface of the foot or hand. Grafts are meshed 1 : 1.5 and secured with tissue glue and veil and dressed with a single layer of Xeroform. The end result will result in improved color match, graft texture, and sensation, less hypertrophic scarring, and improved functionality.

FACE

The face has a crucial functional, cosmetic, and psychological role and facial burns are therefore mostly treated conservatively to allow for maximal tissue preservation, based on the rich capillary network in the face. For superficial burns, removal of blisters and loose debris, followed by the application of a biological skin substitute or topical antiseptics, will yield excellent results. For wounds not healed within 14–21 days, careful tangential excision is performed, preserving as much specialized tissue as possible with formal esthetic reconstructive strategies left for later. Application of allografts is recommended for reassessment if there is doubt about the viability of the recipient area. Grafts should be procured from above the “blush line” if possible.

Similarly, ear burns are treated conservatively with topical antiseptics.

PERINEUM AND GENITALIA

Burns to the perineum and genitalia are usually treated conservatively. However, with continuous fecal soiling, a temporary colostomy should be considered. Full-thickness burns are rare, necessitating a practical approach while awaiting spontaneous healing. Once demarcation has been established, the remaining non-healed areas should be grafted. Contractures can be corrected by scar excision, grafts, or local flaps.

MORTALITY

The *Modified Baux Score* (age × TBSA + 17 for inhalation injury) is a convenient method to determine survival of burn patients as age, TBSA, inhalation injury, and pre-existing comorbidities are still the four main determinants of burn mortality. That said, mortality has decreased to such an extent that most children should be considered as candidates for survival, regardless of age, burn size, and delay in resuscitation or presence of inhalation injury on initial presentation. In a modern pediatric burn care setting, a burn size of approximately 60% TBSA is a crucial threshold for post-burn morbidity and mortality. However, with modern surgical techniques including micrografting techniques, a LD^{50} of 75–80% TBSA is possible, even in a resource-poor environment.

FURTHER READING

Duke JM, Rea S, Boyd JH, et al. Mortality after burn injury in children: A 33-year population-based study. *Pediatrics* 2015; 135: e903–10.

Kamolz LP, Lumenta DB, Parvizi D, et al. Smartphones and burn size estimation: Rapid Burn Assessor. *Ann Burns Fire Disasters* 2014; 27: 101–4.

Kraft R, Herndon DN, Al-Mousawi AM, et al. Burn size and survival probability in paediatric patients in modern burn care: A prospective observational cohort study. *Lancet* 2012; 379(9820): 1013–21.

Rode H, Martinez R, Potgieter D, et al. Experience and outcomes of micrografting for major paediatric burns. *Burns* 2017; 43: 1103–10.

Sterling JP, Heimbach DM. Hemostasis in burn surgery – A review. *Burns* 2011; 37: 559–65.

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Thoracic trauma

JEROME LOVELAND AND NIRAV PATEL

BACKGROUND AND STATISTICS

Trauma is the leading cause of death and disability among children worldwide and accounts for 40% of all childhood mortality in some countries (WHO, 2006). With an estimated annual 6500 deaths in South Africa, trauma is the leading cause of death among children aged 1–14 years. In fact, rates of traumatic injury in South African children are estimated to be between 10 and 25 times those encountered in Europe. Globally, approximately 1 million children die from trauma every year. Although thoracic trauma in children is a relatively rare occurrence, accounting for 5–12% of all pediatric trauma-related admissions, it is associated with significant morbidity and mortality. Unsurprisingly, children who have sustained thoracic trauma are noted to have higher trauma and injury severity scores (TS and ISS) than those without thoracic trauma. In isolation, thoracic trauma accounts for about 5% of pediatric trauma-related mortalities. That said, at least one-third of all thoracic injuries are associated with significant injury to other systems, typically involving the head and abdomen. In these instances, mortality increases to about 40%.

According to available data, blunt-force trauma accounts for between 85% and 90% of thoracic trauma. With the increasing use of firearms and knives, penetrating thoracic injury is becoming a more common occurrence, especially in male children. Nearly two-thirds of recorded thoracic trauma occurs in boys, with road traffic injuries (motor vehicle and pedestrian) and falls from height accounting for the majority of injuries.

Lung contusions, rib fractures, and pneumo/hemothorax account for the overwhelming majority of thoracic injuries. Injuries to the diaphragm and mediastinal structures including the heart, aorta, and other great vessels, tracheobronchial tree, and esophagus are much less common, but these are associated with significantly higher morbidity and mortality.

ANATOMY AND PHYSIOLOGY

Any attempt to understand pediatric thoracic trauma must take into account some important principles:

- The pattern of trauma varies according to the age of the affected child. For instance, due to the high pliability of the chest wall and relative deficiency of muscle in young children, significant pleural and parenchymal injury is possible without evidence of rib fracture.
- Pediatric airway differences include the relatively larger tongue and softer palate, shorter and narrower trachea, and larger occiput, predisposing the neck to flexion and occlusion of the airway.
- A child has a thinner and more compliant chest wall.
- Lower functional residual capacity and higher oxygen consumption predispose children to hypoxia.
- Increased mobility of the mediastinum predisposes children to obstructive shock from tension pneumothorax.

Injury classification and management

Thoracic trauma may be classified according to mechanism of injury, simply either blunt or penetrating. However, given that the predominant mechanism of injury is blunt, pediatric thoracic trauma may be more appropriately classified anatomically as trauma to the chest wall, pleural cavity or pulmonary parenchyma, and mediastinal structures. Relevant injuries by anatomical location are presented in **Table 83.1** and discussed below.

Table 83.1 Thoracic injury classified by location

Location	Thoracic injury
Chest wall	Rib fracture Flail chest Sucking/open pneumothorax Traumatic asphyxia
Pleural cavity/lung parenchyma	Lung contusion Pneumothorax/hemothorax Tension pneumothorax Diaphragmatic rupture
Mediastinal structures	Pericardial tamponade Esophageal rupture Cardiac contusion Tracheobronchial injury Great vessel injury

Management of trauma in all children should always follow Advanced Trauma Life Support® (ATLS) protocols. Associated injuries are still the most important predictors of morbidity and mortality in pediatric thoracic trauma. As such, rigorous primary and secondary surveys and concomitant treatment of associated injuries are critical to the management of childhood thoracic injury.

CHEST WALL

Rib fractures

Due to the high compliance of the chest wall in young children, rib fractures are infrequent in children under the age of 8 years. In fact, rib fractures only occur in 2% of all general pediatric trauma cases, although this increases to 60% in cases of thoracic trauma. As the number of rib fractures increases, so too do patients' overall ISS and the likelihood of multisystem injury.

Rib fractures are initially suspected on physical examination, but are most commonly diagnosed on chest X-ray (CXR). In children under 3 years of age, rib fractures should raise suspicion of non-accidental injury and appropriate further investigation and management should be instituted. Fractures of the first and second ribs have traditionally been associated with significant thoracic trauma and should also raise suspicion of injury to mediastinal organs, for instance cardiac contusion or major vascular injury.

Management of isolated rib fractures in children is usually supportive and includes appropriate analgesia, physiotherapy, and, where necessary, supplemental oxygen. Pain may be managed with non-steroidal anti-inflammatory drugs (NSAIDs), opioids, local anesthesia in the form of nerve blocks, and regional anesthesia in the form of thoracic epidurals. In cases of severe respiratory compromise, mechanical ventilation may be considered.

Flail chest

Due to the high compliance of the chest wall in pediatric patients, multiple rib fractures that occur in parallel (flail chest) are an extremely rare occurrence. In fact, flail chest is the least common of all described pediatric thoracic injuries. When present, flail chest is associated with significant injury to the underlying lung thus aggravating respiratory compromise, and may present with paradoxical movement of the affected area of the chest during inspiration and expiration.

As is the case with rib fracture, treatment of flail chest is aimed at management of respiratory depression. Non-invasive management includes analgesia, physiotherapy, supplemental oxygen, and continuous positive end-expiratory pressure. Operative intervention for flail chest is not common, but it may be indicated in instances of severe chest wall deformity and failure to wean mechanical ventilation.

Sucking or open pneumothorax

Sucking or open pneumothorax is a serious but rare condition in children. It is usually associated with blast injury or impalement, and results from a one-way valve being created by the soft tissues of the chest wall. During inhalation, negative pressure in the thoracic cavity results in air being sucked into the chest. Due to the one-way valve effect, this air cannot be expelled on expiration. Consequent progressive accumulation of air within the thorax may rapidly result in collapse of the ipsilateral lung, mediastinal shift, and obstructive shock.

Treatment of an open pneumothorax includes adequate analgesia, supplementary oxygen, insertion of an intercostal drain (ICD), and placement of a dressing that completely occludes the wound to prevent further air from being sucked into the chest. If an ICD is not available, a three-sided dressing is applied.

Traumatic asphyxia

Traumatic asphyxia almost exclusively affects children and occurs when there is sudden compression of the chest against a closed glottis. The resultant increase in intrathoracic pressure is transmitted to the venules and capillaries of the head, neck, and chest and results in their rupture. Traumatic asphyxia may present with conjunctival hemorrhage, facial swelling, and petechial hemorrhage of the face, neck, and chest (Perthe's syndrome) that usually resolve spontaneously. Severe cases of traumatic asphyxia may result in permanent neurological deficit, but the greatest morbidity and mortality from traumatic asphyxia is usually due to associated injuries.

LUNGS AND PLEURAL CAVITY

Injuries to the lung or pleural cavity are the most common sequelae of blunt and penetrating thoracic trauma.

Lung contusion

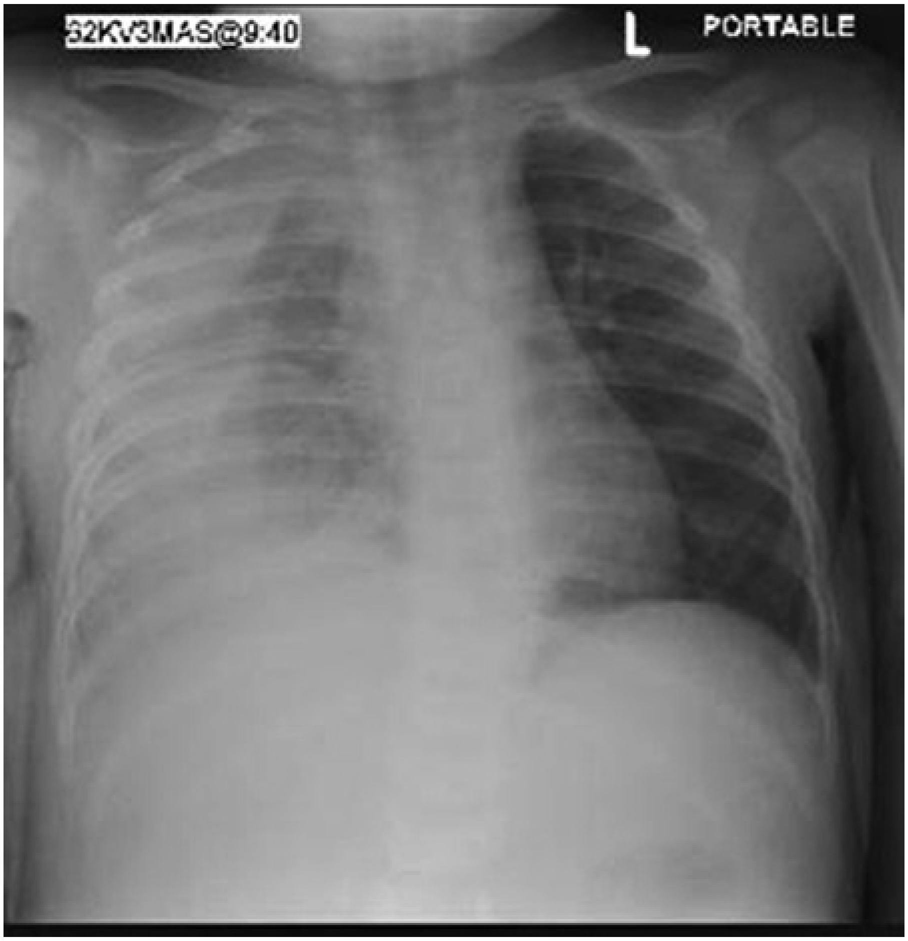
Lung contusion is the most common thoracic injury in children and occurs in about half of all cases of blunt thoracic trauma. It rarely results in respiratory failure requiring mechanical ventilation, and usually resolves within a week of occurrence without scarring of the lung. Pulmonary contusions are usually easily diagnosed on CXR, and may be complicated by pneumo- or hemothorax, respiratory failure, or secondary infection. The most common features of lung contusion on CXR are isolated or confluent opacification in a sporadic distribution.

Lung contusions are typically managed conservatively with supplementary oxygen, analgesia, and physiotherapy, although, if significant hypoxia persists, patients may require a period of ventilation.

Pneumothorax and hemothorax

Pneumothorax and hemothorax are the second most common thoracic injuries. In about two-thirds of cases, these injuries are associated with significant additional thoracic and non-thoracic injury. Pneumothorax may be due to injury to the chest wall, lung parenchyma, tracheobronchial tree, or esophagus. Pneumothorax and hemothorax may be asymptomatic or present with respiratory distress, chest pain, failure to maintain oxygen saturation, tenderness of the chest wall, crepitus from subcutaneous emphysema, decreased air entry on the affected side, and tracheal deviation to the unaffected side. Simple pneumothorax has the propensity to progress to tension pneumothorax due to the relatively mobile mediastinum in children, while a large hemothorax may not be immediately recognized and result in hemorrhagic shock. This is especially true in younger children in whom the clinical features of hypotension are often masked until the time of catastrophic decompensation. It is important to note that each hemithorax may hold up to 40% of a child's total blood volume.

CXR and emergency room ultrasound are the investigations of choice for pneumo- and hemothorax (**Figures 83.1 and 83.2**). Pneumothorax is evidenced by hyperlucency of the affected side, mediastinal shift, a sharply delineated heart border, and a well-defined lung border. Hemothorax may be more difficult to diagnose, especially in the supine position in which most CXRs are taken in resuscitation bays, where blunting of the costophrenic angle that typically demonstrates hemothorax is replaced by so-called "veiling" of the lung as blood is distributed in the posterior hemithorax.

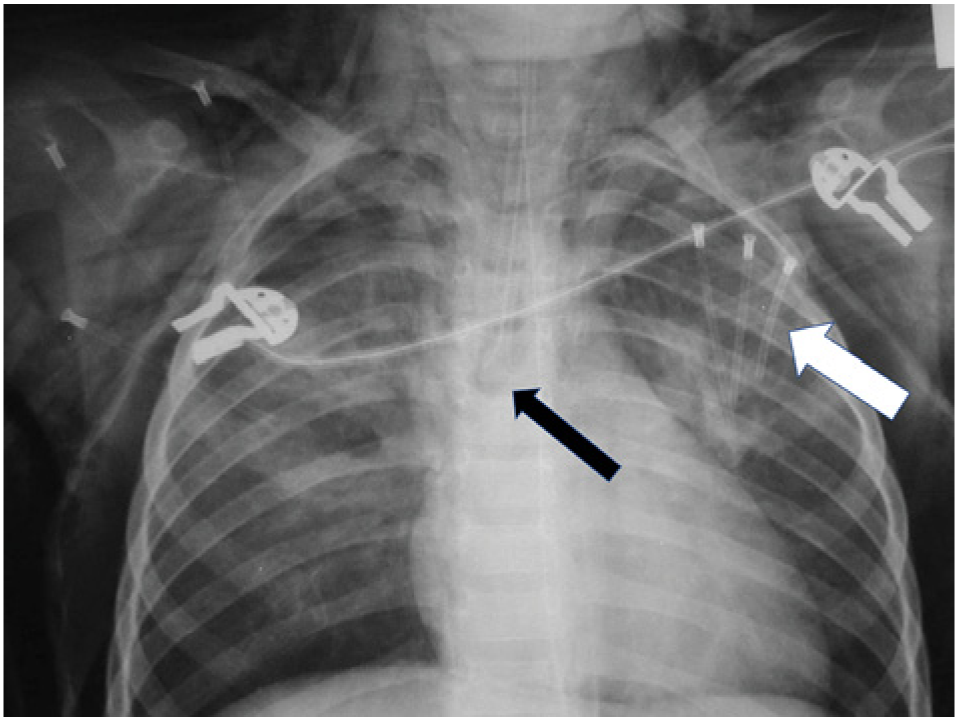


83.1 Right-sided hemothorax after blunt thoracic injury before tube thoracostomy.

Immediate ICD is the treatment of choice for all symptomatic pneumo- and hemothoraces. Asymptomatic pneumothoraces that are not evident on CXR but manifest on computed tomography (CT) scan do not necessarily require drainage. Nevertheless, these patients must be carefully observed in hospital and should be maintained on high-flow oxygen for 24–48 hours. There is some controversy around the conservative management of small-volume hemothoraces. In our experience, all traumatic hemothoraces require drainage by ICD insertion in order to prevent the complications of a retained hemothorax, including pneumonia, atelectasis, lung entrapment, and empyema. Acute, massive pulmonary hemorrhage may be rapidly addressed through median sternotomy and hilar clamping. Such a maneuver stems bleeding from the lung parenchyma and affords the opportunity to determine if the injury may be optimally addressed through a non-anatomical resection of lung, lobectomy, or pneumonectomy.

Diaphragmatic rupture

Diaphragmatic injury in children is usually secondary to high-energy blunt force trauma and has a strong association with multisystem injury. Injury is evidenced by the presence of intrathoracic hollow viscera either on CXR (**Figure 83.2**) or CT scan of the chest and is most commonly noted on the left due to the protective presence of the liver on the right.



83.2 Overzealous needle decompression (white arrow) due to suspected bilateral pneumothoraces. Decreased air entry actually noted to be secondary to inadvertent intubation of the right main bronchus intubation (black arrow) on the CXR.

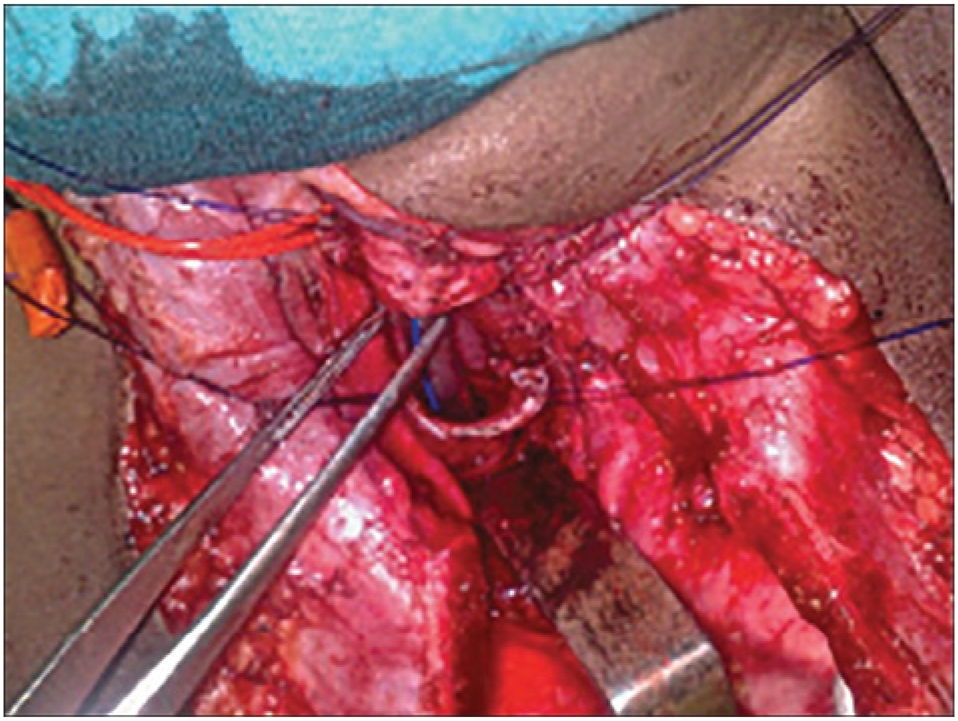
Operative management of diaphragmatic injury is usually by open laparotomy in order to address associated intra-abdominal injuries, for instance splenic and liver laceration, or bowel perforation.

MEDIASTINAL STRUCTURES

Injuries to mediastinal structures are the least common of all blunt and penetrating thoracic traumas. However, these injuries carry the greatest morbidity and mortality. Mediastinal injury may be categorized as injury to the tracheobronchial tree, injury to the esophagus, or injury to the heart and great vessels.

Tracheobronchial injury

Airway trauma in childhood is most commonly due to high acceleration–deceleration forces and usually occurs within the membranous portion of the trachea 2–3 cm from the carina (**Figure 83.3**). With respect to major bronchial injuries, approximately half occur within the proximal 2 cm of the right main stem bronchus. In the majority of cases, patients with proximal airway injuries present with respiratory distress, stridor, cyanosis, hemoptysis, chest pain, and massive subcutaneous emphysema. In cases of more distal airway disruption, patients may present with features of pneumothorax or tension pneumothorax. Persistent pneumothorax or pneumomediastinum, or a constantly bubbling chest drain, are features of major airway injury and should prompt further investigation by rigid or flexible bronchoscopy or chest CT.



83.3 Access to the trachea provided by median sternotomy incision showing lacerated tracheal injury.

Significant airway injury usually requires surgical exploration, via a posterolateral thoracotomy or sternotomy. Proximal lacerations and disruptions are most often treated by direct repair, while distal lacerations are typically managed by direct repair, and in some cases non-anatomical pulmonary resection. Non-operative management of airway injury is possible in cases where less than one-third of the circumference of the bronchi is lacerated, but it is often associated with airway stenosis and its long-term sequelae (i.e. chronic infection and bronchiectasis).

Esophageal injury

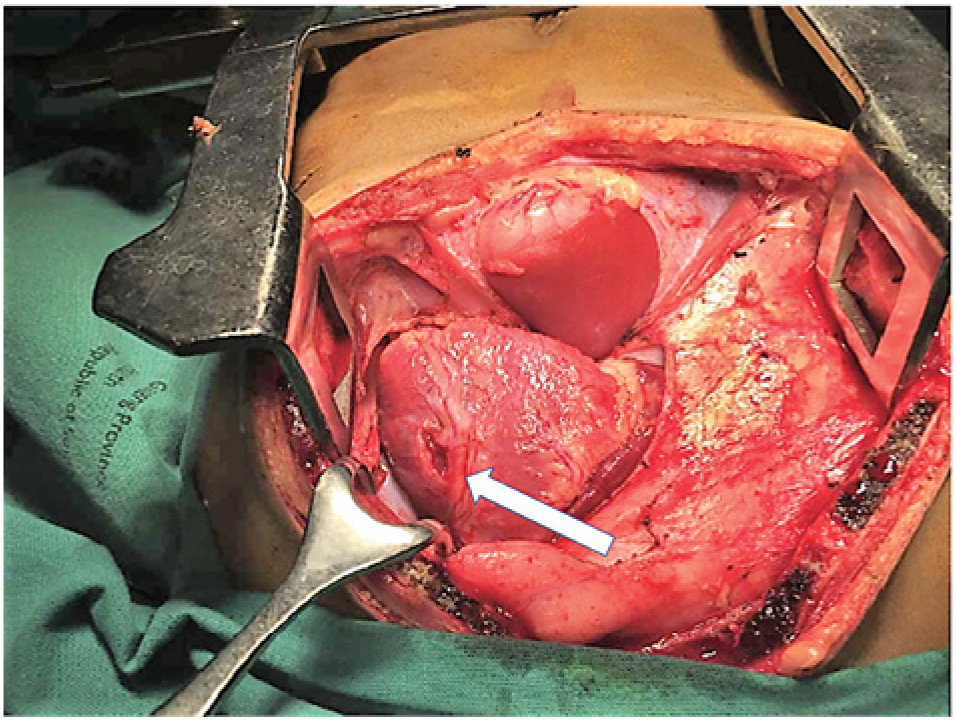
Penetrating injury is the most common cause of esophageal trauma in children. Given the non-specific presentation of esophageal injury, typical signs and symptoms including fever, chest pain, dysphagia, subcutaneous emphysema, and hydrothorax, and the extremely high morbidity and mortality associated with mediastinitis, clinicians should maintain an extremely high index of suspicion for esophageal injury. When suspected, injury should be confirmed by contrast study, endoscopy, or contrast-enhanced CT scan. As per adult literature, accurate diagnosis of esophageal injury increases significantly when endoscopy is used in conjunction with a contrast study.

The management of esophageal injury is largely dependent on the timing of the injury and underlying status of the patient. For relatively "fresh" injuries, typically defined as those occurring within 24 hours of presentation, primary repair is indicated. This repair may constitute direct repair of the esophagus in a single layer, or direct repair with pleural flap cover. For those patients presenting more than 24–48 hours after injury, operative repair is not indicated and treatment should be directed toward the management of mediastinitis by means of mediastinal drainage, IV antibiotics, and parenteral nutrition. In cases of severe injury complicated by mediastinitis, management may also include esophageal diversion in order to limit ongoing mediastinal contamination.

Cardiac and great vessel injury

Blunt trauma to the heart may result in various injuries, including myocardial concussion, myocardial contusion, myocardial rupture, or septal or valvular rupture. These injuries may present with sudden death, changes in cardiac rhythm as evidenced on electrocardiogram (ECG), onset of a new cardiac murmur, or features of restrictive or cardiogenic shock. *Beck's triad* is a combination of diminished blood pressure, muffled heart sounds on auscultation, and jugular venous distension implying fluid, in this case blood, in the pericardial sac. While specific discussion of these injuries is beyond the scope of this text, it is important for clinicians to be suspicious of cardiac injury. Appropriate investigation includes CXR, ECG, echocardiography, and cardiac enzymes.

Appropriate management of cardiac trauma depends on the specific nature of the cardiac injury, as well as the severity and extent of associated injuries. Suspected penetrating injury to the heart mandates urgent sternotomy and repair (**Figure 83.4**).



83.4 Stab wound (white arrow) of the left ventricle medial to the left anterior descending artery.

Blunt aortic injury occurring in the thoracic aorta is the most common aortic injury in children. These injuries are most often the result of acceleration–deceleration forces encountered in high-velocity road traffic accidents (pedestrian and motor vehicle). Suspicion for aortic injury should always be high, particularly in hemodynamically unstable patients, after blunt thoracic trauma as external evidence of great vessel injury is often limited. A widened cardiothoracic ratio, straightening of the aortic knuckle, right-sided tracheal diversion, and depression of the left main bronchus on CXR are highly suggestive signs of great vessel injury and warrant further investigation by CT angiography. On CT angiography, the most commonly detected lesions are pseudoaneurysms within the descending aorta. Unfortunately, few studies on blunt aortic injury in children exist, though available data do demonstrate that up to 70% of those children that survive to diagnosis, survive their underlying injuries.

Management of thoracic aortic injuries may be open or radiological, although this is dependent on numerous factors including the services and skills available at a particular center, the specific nature of the aortic injury, and the general condition of the patient.

SURGICAL EXPOSURE AND INTERVENTIONS

Insertion of an intercostal drain

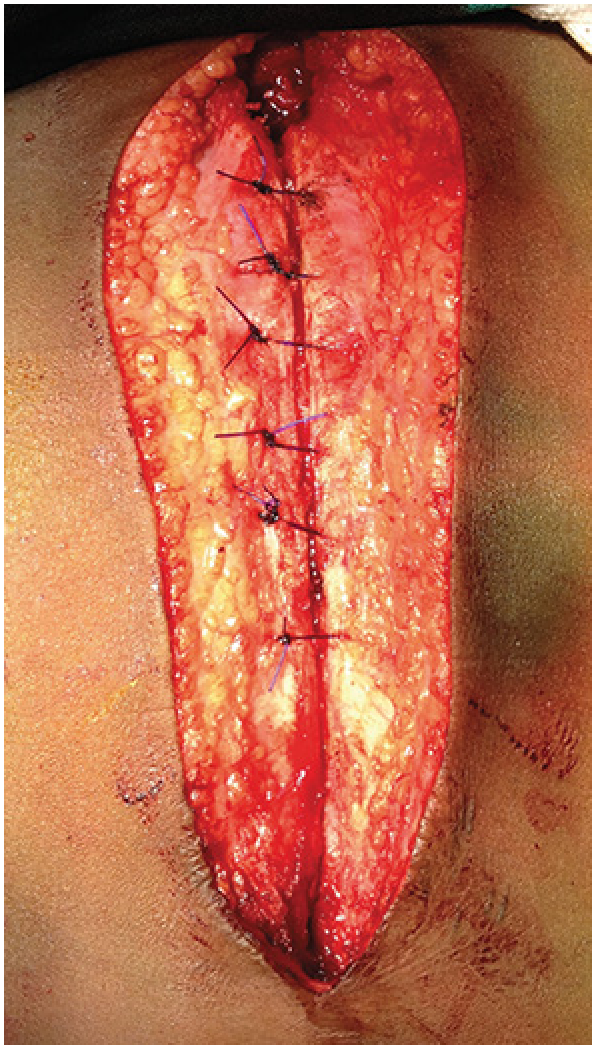
ICD insertion in children follows the same principles and critical steps as in adults. That said, allowance must be made for adequate and safe sedation and analgesia, appropriate positioning of the child during tube thoracostomy, anatomical factors including the smaller size of the triangle of safety (zone incorporating the area between the third and fifth intercostal space between the mid and anterior axillary lines) and thinner chest wall, and appropriate securing of the ICD once in place.

Surgical approaches

Exposure of thoracic and mediastinal organs may be achieved through a multitude of well-described surgical approaches including anterolateral thoracotomy, posterolateral thoracotomy, clamshell thoracotomy, and median sternotomy. Though well described in the adult population, such interventions are rarely necessary in the management of life-threatening blunt or penetrating thoracic trauma in children, with the majority of patients responding to simple ICD. Possible indications for surgical exploration include, but are not limited to:

- Ongoing thoracic bleeding in spite of ICD, e.g. bleeding equivalent to 2–3 mL/kg/hour over 3 hours
- Esophageal disruption
- Tracheobronchial disruption
- Diaphragmatic injury
- Penetrating cardiac injury

Median sternotomy is the preferred approach for emergent thoracic surgical exploration in our unit. This approach is extremely well tolerated, enables quick, easy, and wide access, and can easily be extended along the clavicles or into the neck or abdomen, as required. In young children the sternum is easily divided with a pair of sturdy scissors or Gigli wire; in older children this may be quickly achieved with a pneumatic saw or sternal blade. Further advantages of this approach include the absence of injury to intercostal muscles and the ribs, lower postoperative analgesic and ventilatory requirements than thoracotomy (in our experience), ease of closure with PDS sutures (**Figure 83.5**), and prevention of potentially disfiguring damage to a child's breast or pectoralis muscle.



83.5 Closed median sternotomy in a child.

Median sternotomy is achieved through an incision extending from the suprasternal notch to the xiphisternum. The sternum is divided and the right and left hemithoraces exposed, offering immediate access to the anterior and posterior pericardium, right and left lungs and pulmonary hila, trachea and esophagus, and diaphragm.

Resuscitative thoracotomy for children presenting *in extremis* may be achieved through a left anterolateral thoracotomy in the fifth intercostal space. Such exposure facilitates immediate access to the posterior pericardium and the intrathoracic aorta. In instances, where further exposure is required and a resuscitative thoracotomy has been performed, the incision may be extended across the sternum into the right chest, the so-called *"clamshell" thoracotomy*. In our experience, median sternotomy facilitates equivalent exposure, may be achieved as rapidly, and is associated with lower morbidity than clamshell thoracotomy.

CONCLUSION

Though relatively uncommon, thoracic trauma in children is associated with multisystem injury and significant morbidity and mortality. Clinicians should always maintain a high index of suspicion when assessing children with potential for thoracic trauma due to the potential for life-threatening thoracic and non-thoracic injury. There is ongoing debate on the role of CT scan in the diagnosis of thoracic trauma, and the existing body of evidence does not support the routine use of CT scanning in the workup of the patient, particularly in the presence of a normal CXR.

Immediate operative intervention is rarely indicated in pediatric thoracic trauma and should be reserved for instances of major tracheobronchial injury, great vessel injury, cardiac injury requiring repair or drainage, and esophageal perforation. When indicated, such intervention is best undertaken through median sternotomy.

FURTHER READING

Herbert HH, van As AB, Bachani AM, et al. Patterns of pediatric injury in South Africa: An analysis of hospital data between 1997 and 2006. *J Trauma Acute Care Surg* 2012; 73: 168–74.

Pearson EG, Fitzgerald CA, Santore MT. Pediatric thoracic trauma: Current trends. *Semin Pediatr Surg* 2017; 26: 36–42.

Ruiz-Elizalde A, Tuggle DW. Thoracic trauma. In: Holcomb GW, Murphy PJ, Ostlie DJ (eds). *Ashcraft's Pediatric Surgery*, 6th ed. Philadelphia: Elsevier; 2014: pp. 190–7.

van As AB, van Nieker A. Prevention of childhood injuries. *S Afr Med J* 2017; 107: 182.

Wesson DE, Cox CS. Thoracic injuries. In: Coran A, Scott Adzick N, Krummel T, et al. (eds). *Pediatric Surgery*, 7th ed. Philadelphia: Elsevier; 2012: pp. 271–87.

World Health Organization (WHO). Child and Adolescent Injury Prevention: A WHO Plan of Action 2006–2015. Geneva: World Health Organization, 2006.

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Abdominal trauma

STEVEN STYLIANOS AND MARK V. MAZZIOTTI

HISTORY

Who could have imagined the influence of James Simpson's publication in 1968 on the successful non-operative treatment of select children presumed to have splenic injury? Initially suggested in the early 1950s by Tim Warnsborough, then Chief of General Surgery at the Hospital for Sick Children in Toronto, it is remarkable to consider that the era of non-operative management for pediatric spleen injury began with the report of 12 children treated between 1956 and 1965. The diagnosis of splenic injury in this select group was made by clinical findings together with routine laboratory and plain X-ray findings. It should be borne in mind that the report predated ultrasound (US), computed tomography (CT), or isotope imaging.

Nearly half a century later, the standard treatment of hemodynamically stable children with splenic injury is non-operative, and this concept has now been successfully applied to most blunt injuries of the liver, kidney, and pancreas. Surgical restraint has been the theme, based on an increased awareness of the anatomic patterns and physiologic responses characteristic of injured children. Our colleagues in adult trauma care have slowly acknowledged this success and applied many of the principles learned in pediatric trauma to their patients.

Few surgeons have extensive experience with massive abdominal solid organ injury requiring immediate surgery. It is imperative that surgeons familiarize themselves with current treatment algorithms for life-threatening abdominal trauma. Important contributions have been made to the diagnosis and treatment of children with abdominal injury by radiologists and endoscopists. The resolution and speed of CT, the screening capabilities of focused abdominal sonography for trauma (FAST), and the percutaneous, angiographic, and endoscopic interventions of non-surgeon members of the pediatric trauma team have all enhanced patient care and improved outcomes. This chapter focuses on the more common blunt injuries of the spleen, liver, duodenum, pancreas, and kidney.

RESUSCITATION

A multitrauma patient entering a medical facility should be treated by an organized team of surgeons, physicians, and nurses. The composition of the team will vary, but the senior surgeon should be the team leader. Preparation is mandatory and includes ensuring the availability of equipment appropriate to children of varying ages and the establishment of a resuscitation protocol. The “checklist approach” is the surest way to accomplish the essential steps of diagnosis and treatment while individualizing care for each patient. The initial evaluation of the acutely injured child is similar to that of the adult. Plain radiographs of the C-spine (cervical spine), chest, and pelvis are obtained following the initial survey and evaluation of A (airway), B (breathing), and C (circulation). As imaging modalities have improved, treatment algorithms have changed significantly in children with a suspected intra-abdominal injury. Prompt identification of potentially life-threatening injuries is now possible in the vast majority of children.

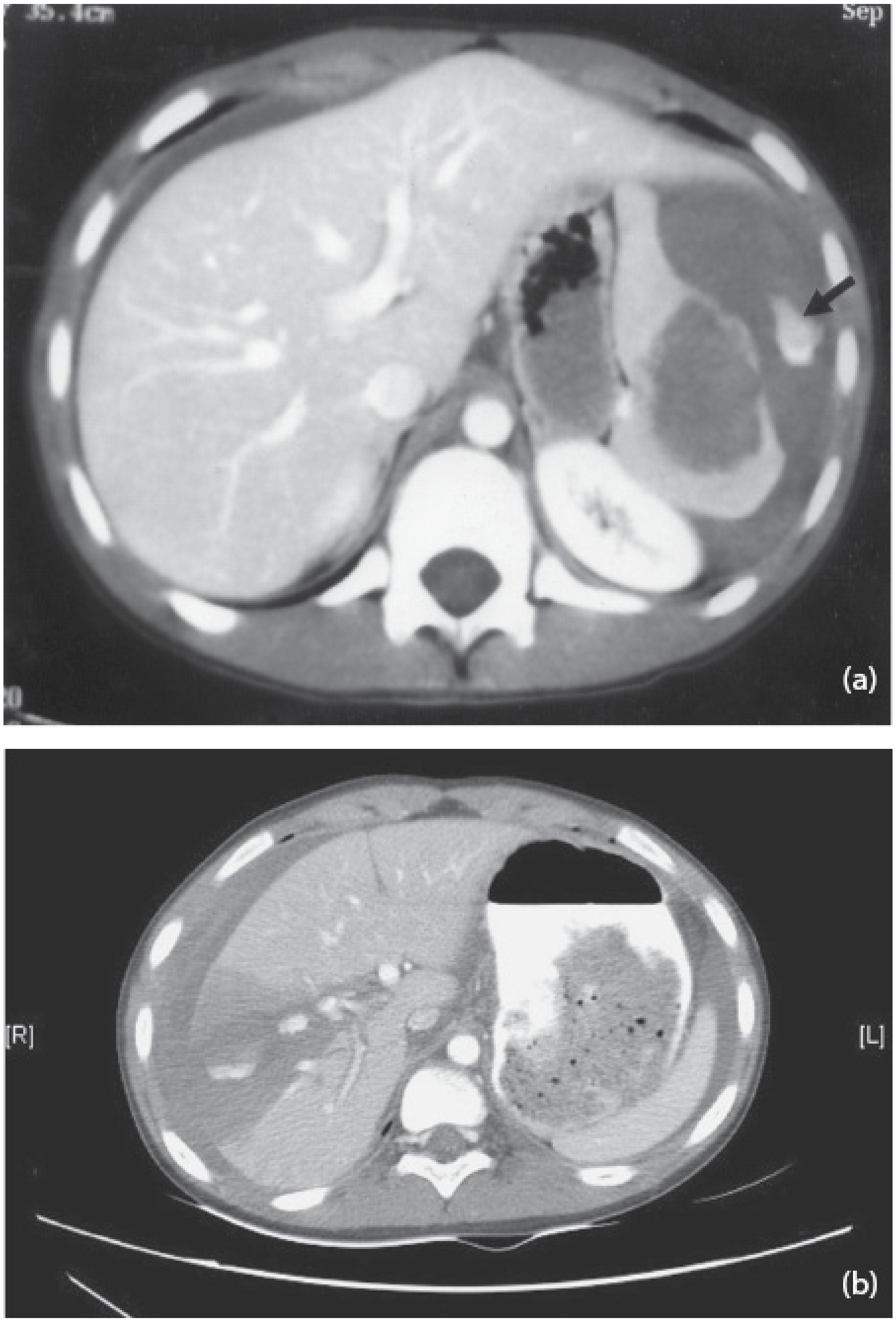
Airway control, vascular access, spinal precautions, and temperature regulation must be assured throughout the initial evaluation and treatment of injured children. The reader is referred to the *Textbook of Pediatric Advanced Life Support* (American Academy of Pediatrics) and the *Textbook of Advanced Trauma Life Support* (American College of Surgeons) for specific details of airway management, pharmacologic therapy, and central venous access in injured children.

DIAGNOSIS OF BLUNT ABDOMINAL INJURIES

Recognition of significant abdominal injuries in children with blunt multisystem trauma can be difficult. Physical examination is inaccurate in more than 30% of cases, particularly in those patients seen soon after injury or with central nervous system injuries and an abnormal neurologic examination. It is no surprise that advances in diagnosis have paralleled the development of new imaging technology. Prompt and accurate recognition of abdominal injuries, now possible with CT, allows for focused treatment plans.

CT has become the imaging study of choice for the evaluation of injured children because of several advantages. It is now readily accessible in most healthcare facilities, is non-invasive, is a very accurate method of identifying and qualifying the extent of abdominal injury, and has reduced the incidence of non-therapeutic exploratory laparotomy. Controversy remains regarding the benefits of enteral contrast for diagnosis of gastrointestinal (GI) tract injuries. Many authors conclude that CT with enteral contrast does not improve diagnosis of GI injuries in the acute trauma setting and can lead to delays in diagnosis and aspiration.

The use of IV contrast is essential, and the utilization of "dynamic" methods of scanning has optimized vascular and parenchymal enhancement (**Figure 84.1a and b**). The impact on resource utilization and outcome of a contrast "blush" on CT in children with blunt spleen and liver injury continues to be debated. Many emphasize that CT blush is worrisome, but that most patients can still be managed successfully without operation. The role and impact of angiographic embolization in adults is still debated and has yet to be determined in pediatric spleen injury. Initial retrospective studies have found angiographic embolization to be safe and effective in children, however, selection criteria remain undefined.

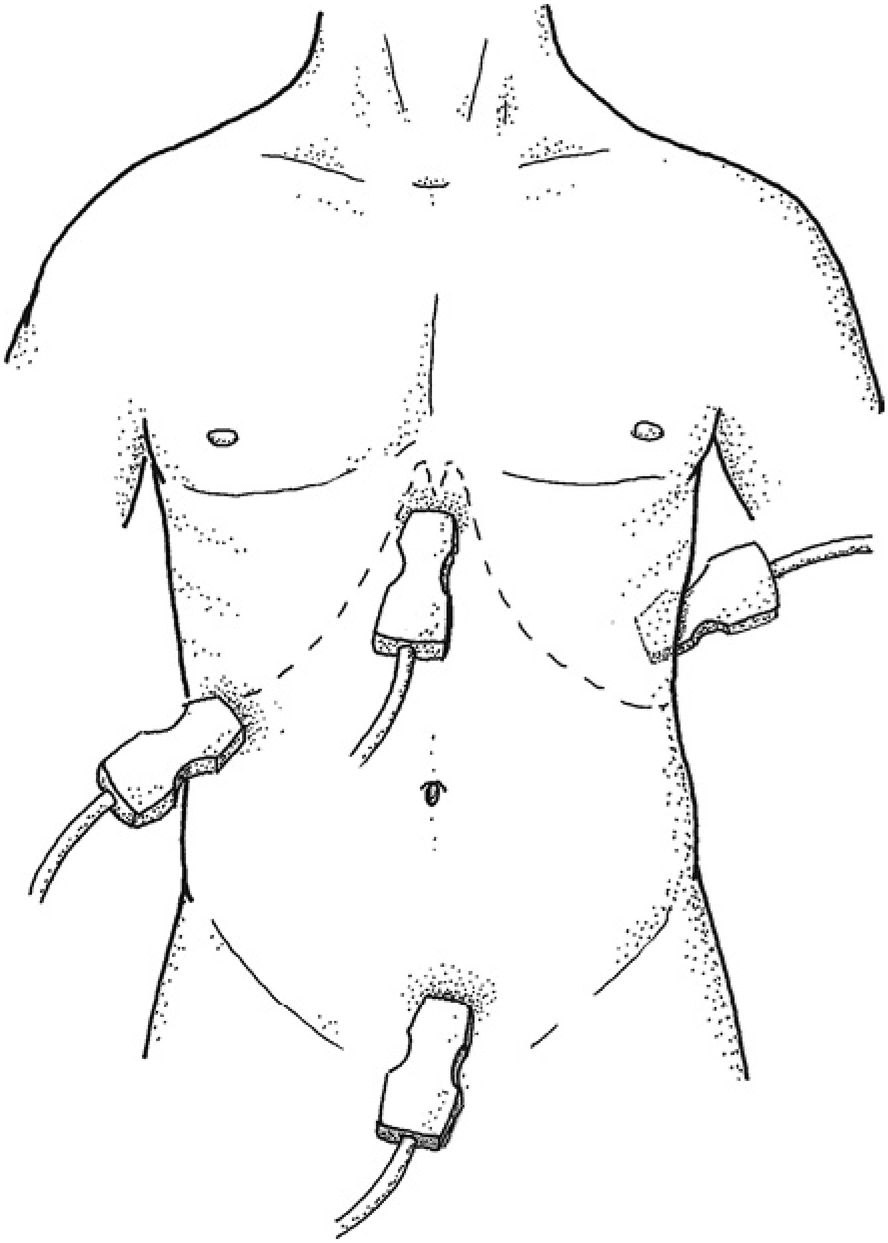


84.1a,b

A head CT, if indicated, should be performed first without contrast, to avoid contrast concealing a hemorrhagic brain injury.

Focused abdominal sonography for trauma

Clinician-performed sonography for the early evaluation of the injured child is currently being assessed to determine its optimal use. The standard four-view FAST examination includes Morrison's pouch, the left flank to include the perisplenic anatomy, and a subxiphoid view to visualize the pericardium and the pouch of Douglas/pelvis (**Figure 84.2**). This bedside examination may be useful as a rapid screening study, particularly in those patients too unstable to undergo an abdominal CT scan. Early reports have found FAST to be a useful screening tool in children, with a high specificity (95%), but a low sensitivity (33%) in identifying intestinal injury. A lack of identifiable free fluid does not exclude a significant injury. FAST may be very useful in decreasing the number of CT scans performed for "low-likelihood" injuries. A recent meta-analysis of FAST in pediatric blunt trauma patients revealed modest sensitivity for hemoperitoneum. The authors concluded that a negative FAST may have questionable utility as the sole diagnostic test to rule out the presence of an intra-abdominal injury. A hemodynamically stable child with a positive FAST should undergo CT.



84.2

Laparoscopy

Recent literature has suggested that laparoscopy can both diagnose and, in some cases, allow definitive surgical management without laparotomy, further limiting the usefulness of diagnostic peritoneal lavage (DPL).

Large series using laparoscopy in adults have demonstrated increased diagnostic accuracy, definitive management of related injuries, decreased non-therapeutic laparotomy rates, and a significant decrease in length of hospital stay, with an attendant reduction in costs. As with elective abdominal surgery, the role of laparoscopy in trauma will increase substantially as trauma centers redirect their training of residents to this modality and as more pediatric centers report outcome studies for laparoscopic trauma management in children.

TREATMENT OF SPECIFIC ABDOMINAL INJURIES

Spleen and liver

The spleen and liver are the organs most commonly injured in blunt abdominal trauma, with each accounting for one-third of injuries. Non-operative treatment of isolated splenic and hepatic injuries in stable children is now standard practice.

The American Pediatric Surgical Association (APSA) Trauma Committee defined consensus guidelines for resource utilization in hemodynamically stable children with isolated liver or spleen injury based on CT grading in 2000. Consensus guidelines on intensive care unit (ICU) stay, length of hospital stay, use of follow-up imaging, and physical activity restriction for clinically stable children with isolated spleen or liver injuries (Grades I–IV) were defined by analysis of this database (**Table 84.1**).

Table 84.1 Proposed guidelines for resource utilization in children with isolated spleen or liver injury

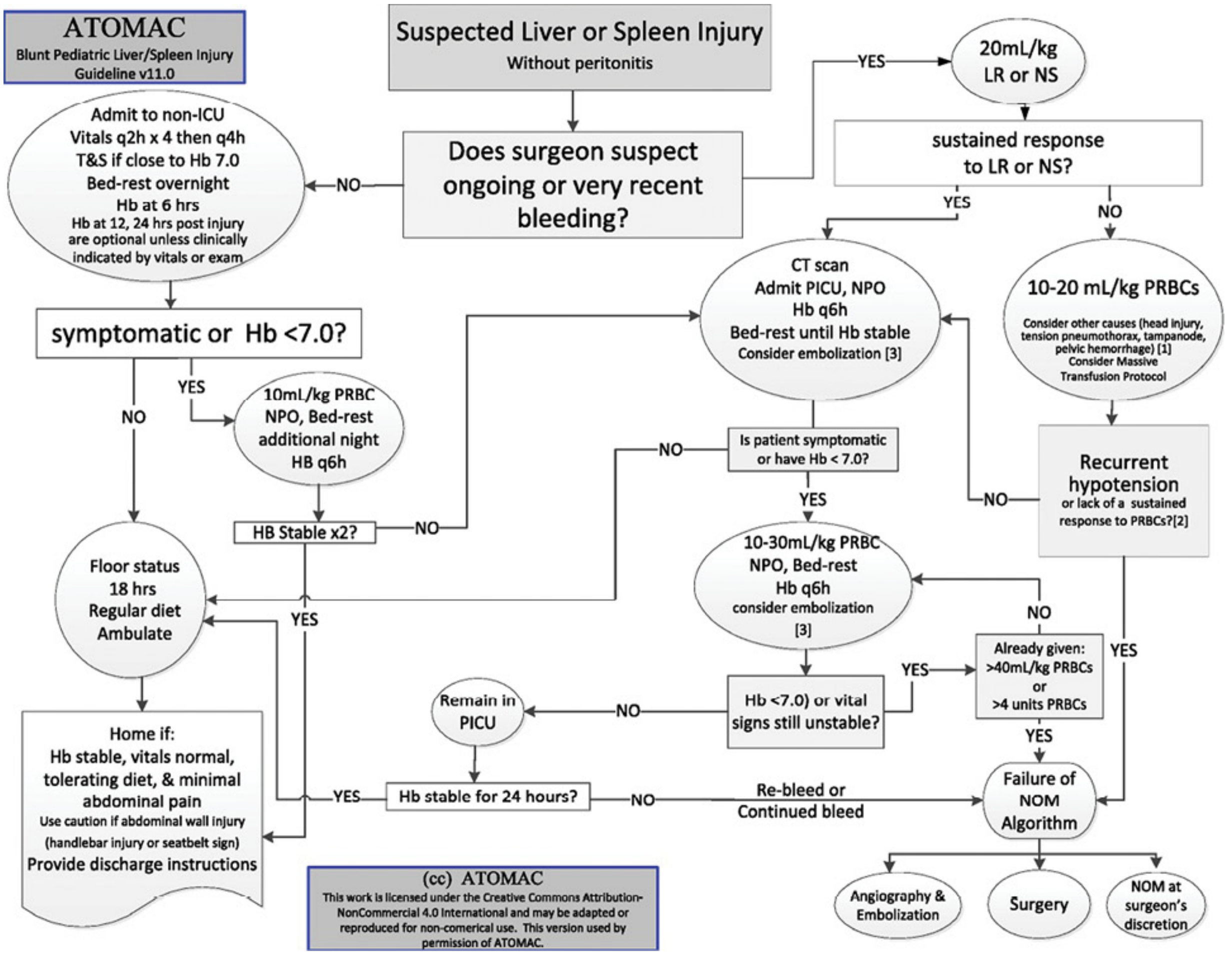
CT grade	I	II	III	IV
ICU days	None	None	None	1
Hospital stay (days)	2	3	4	5
Pre-discharge imaging	None	None	None	None
Post-discharge imaging	None	None	None	None
Activity restriction (weeks)[a]	3	4	5	6

Abbreviations: CT, computed tomography; ICU, intensive care unit.

[a] Return to full contact, competitive sports (i.e. football, wrestling, hockey, lacrosse, mountain climbing, etc.) should be at the discretion of the individual pediatric trauma surgeon. The proposed guidelines for return to unrestricted activity include "normal" age-appropriate activities.

The guidelines were then applied prospectively in 312 children. It is imperative to emphasize that these proposed guidelines assume hemodynamic stability. The extremely low rates of transfusion and operation document the stability of the study patients. Compared with the previously studied 832 patients, the 312 patients managed prospectively by the proposed guidelines had a significant reduction in ICU stay ($p < 0.0001$), hospital stay ($p < 0.0006$), follow-up imaging ($p < 0.0001$), and interval of physical activity restriction ($p < 0.04$) within each grade of injury. The pendulum continues to swing toward less hospitalization in stable children with solid liver or spleen injury. Retrospective and prospective studies have now shown that the APSA guidelines for hospital length of stay can be reduced further.

Authors from the Arkansas Children's Hospital reported on an abbreviated protocol based on hemodynamics while "throwing out" the CT grade of injury in 101 patients with isolated spleen or liver injury. Their protocol resulted in a significant reduction in length of stay (3.5 versus 1.9, $p < 0.001$) from that predicted by APSA guidelines. These observations led to a multi-institutional study and guideline proposal from the Arizona-Texas-Oklahoma-Memphis-Arkansas-Consortium (ATOMAC) (Figure 84.3). The guideline has been validated in over 100 patients with safe and effective care leading to a significant reduction of hospital length of stay.


ATOMAC
Blunt Pediatric Liver/Spleen Injury Guideline v11.0
Suspected Liver or Spleen Injury
Without peritonitis
Does surgeon suspect ongoing or very recent bleeding?
NO
YES
20mL/kg LR or NS
sustained response to LR or NS?
YES
NO
Admit to non-ICU
Vitals q2h x 4 then q4h
T&S if close to Hb 7.0
Bed-rest overnight
Hb at 6 hrs
Hb at 12, 24 hrs post injury are optional unless clinically indicated by vitals or exam
CT scan
Admit PICU, NPO
Hb q6h
Bed-rest until Hb stable
Consider embolization [3]
10-20 mL/kg PRBCs
Consider other causes (head injury, tension pneumothorax, tampanode, pelvic hemorrhage) [1]
Consider Massive Transfusion Protocol
symptomatic or Hb <7.0?
YES
NO
10mL/kg PRBC
NPO, Bed-rest
additional night
HB q6h
Is patient symptomatic or have Hb < 7.0?
NO
YES
Recurrent hypotension
or lack of a sustained response to PRBCs?[2]
NO
HB Stable x2?
NO
YES
Floor status
18 hrs
Regular diet
Ambulate
10-30mL/kg PRBC
NPO, Bed-rest
Hb q6h
consider embolization
[3]
NO
YES
Already given:
>40mL/kg PRBCs
or
>4 units PRBCs
YES
Remain in PICU
NO
Hb <7.0) or vital signs still unstable?
YES
Home if:
Hb stable, vitals normal, tolerating diet, & minimal abdominal pain
Use caution if abdominal wall injury
(handlebar injury or seatbelt sign)
Provide discharge instructions
YES
Hb stable for 24 hours?
NO
Re-bleed or Continued bleed
Failure of NOM Algorithm
Angiography & Embolization
Surgery
NOM at surgeon's discretion
(cc) ATOMAC
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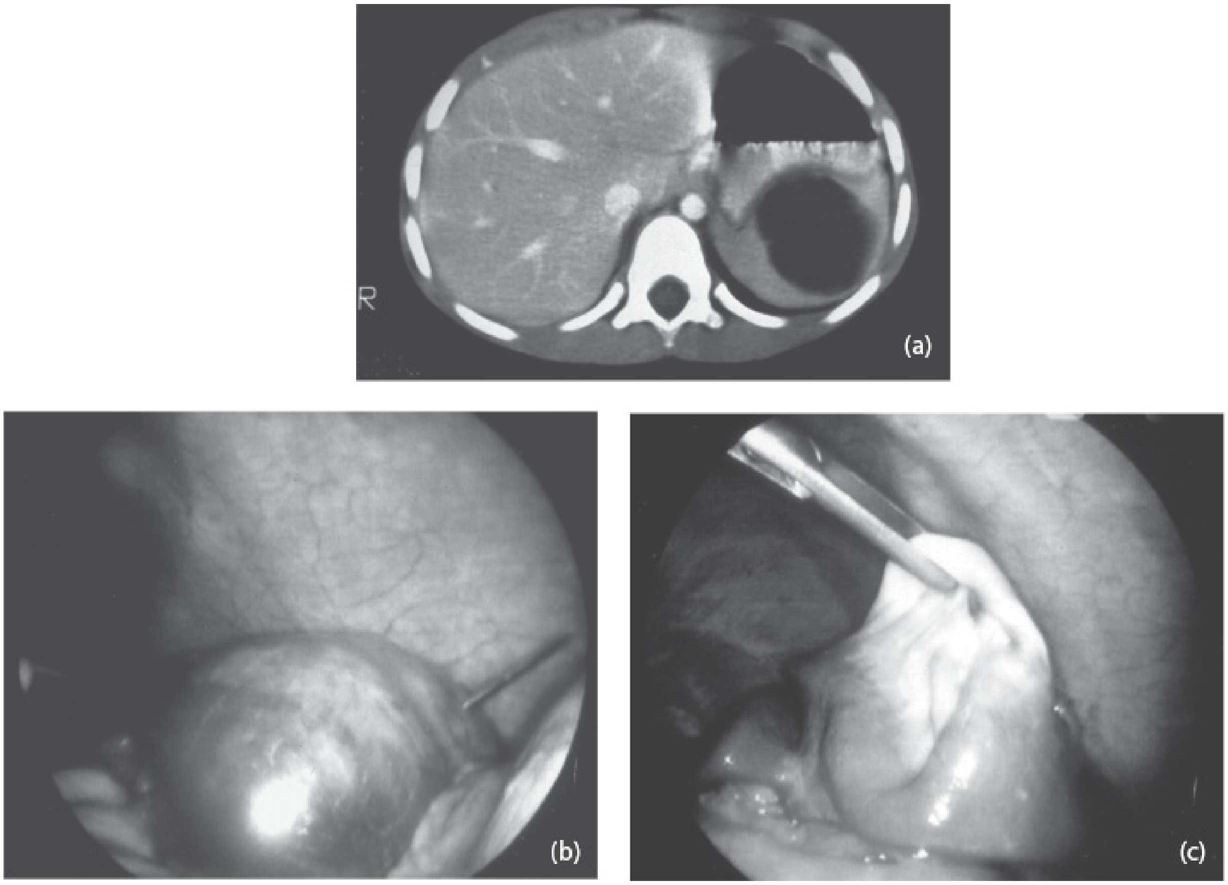
84.3 ATOMAC guideline for management of pediatric blunt liver–spleen injury (LR, lactated Ringer's solution; NOM, non-operative management; NPO, nil per os; NS, normal saline; PICU, pediatric intensive care unit; PRBC, packed red blood cells; T&S, type and screen.) (Modified from Notrica DM, et al. *J Trauma Acute Care Surg* 2015; 79(4): 683–93.)

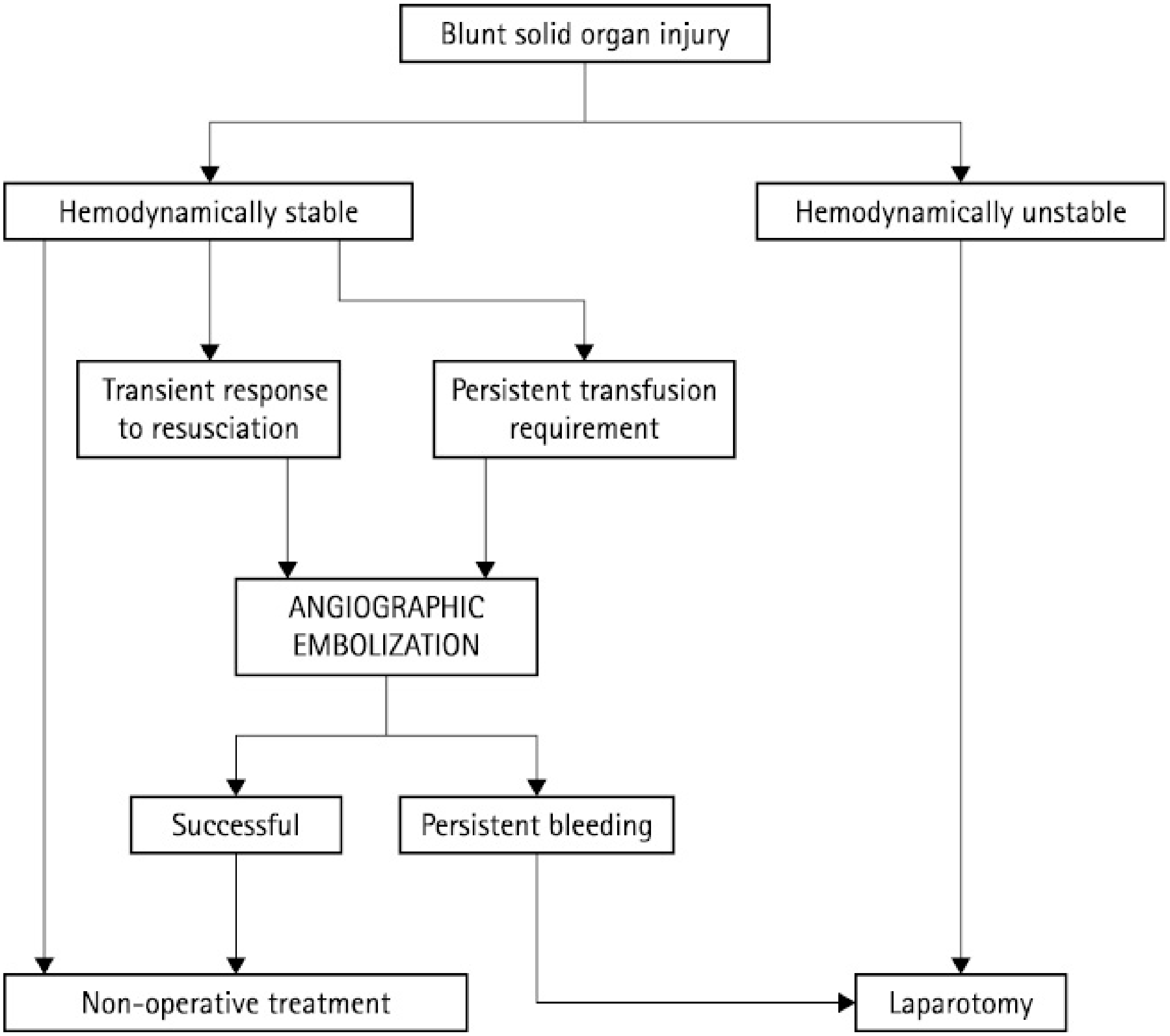
Routine follow-up imaging studies have identified pseudocysts and pseudoaneurysms following splenic injury. Splenic pseudoaneurysms often cause no symptoms and appear to resolve with time. The true incidence of self-limited, post-traumatic splenic pseudoaneurysms is unknown, as routine follow-up imaging after successful non-operative treatment has been largely abandoned. Once identified, the actual risk of splenic pseudoaneurysm rupture is also unclear. These lesions can be treated successfully with angiographic embolization techniques, obviating the need for open surgery and loss of splenic parenchyma.

Splenic pseudocysts can achieve enormous size, leading to pain and GI disturbance (**Figure 84.4a**). Simple percutaneous aspiration leads to a high recurrence rate. Laparoscopic excision and marsupialization is highly effective (**Figure 84.4b and c**).

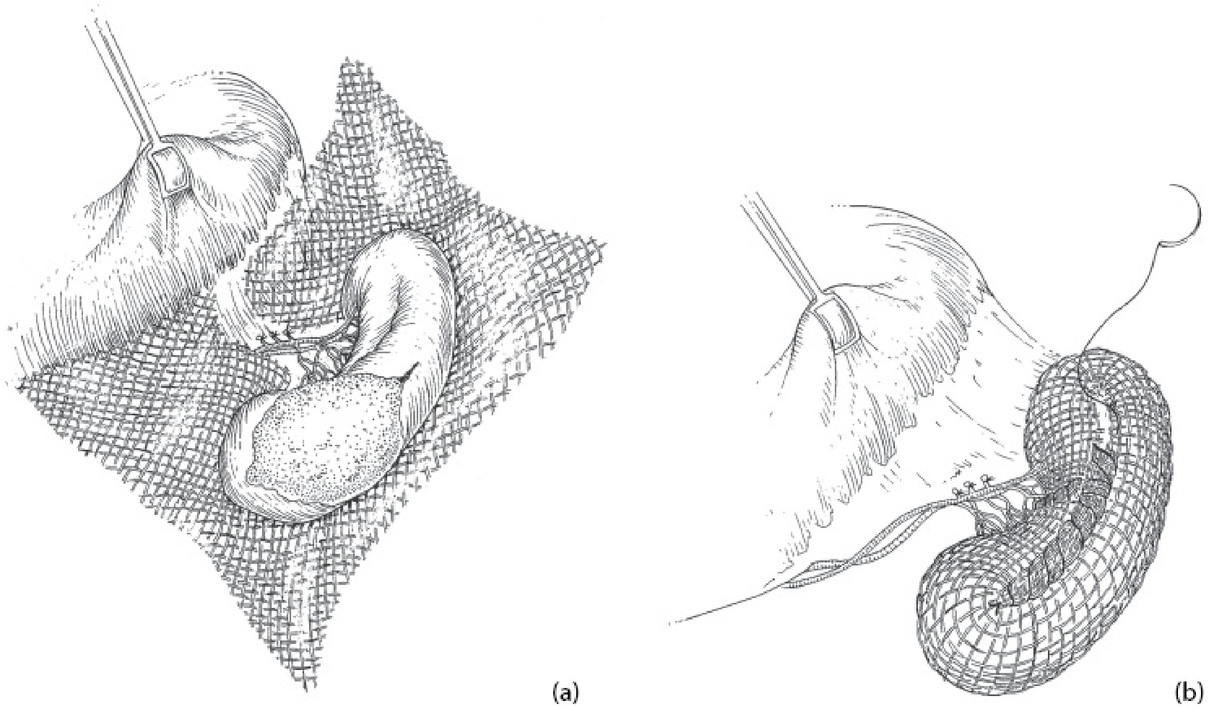


84.4a–c

Even the most severe solid organ injuries can be treated without surgery if there is prompt response to resuscitation. In contrast, emergency laparotomy and/or embolization are indicated in patients who are hemodynamically unstable, despite fluid and red blood cell transfusion (**Figure 84.5**). Fenton et al. evaluated 605 patients with abdominal solid organ injury and found that 18 (3%) had angiography. Embolization of the splenic artery was performed in 11 of 18 patients. Most spleen and liver injuries requiring operation are amenable to simple methods of hemostasis using a combination of manual compression, direct suture, topical hemostatic agents, and woven polyglycolic mesh wrapping (**Figure 84.6a and b**).

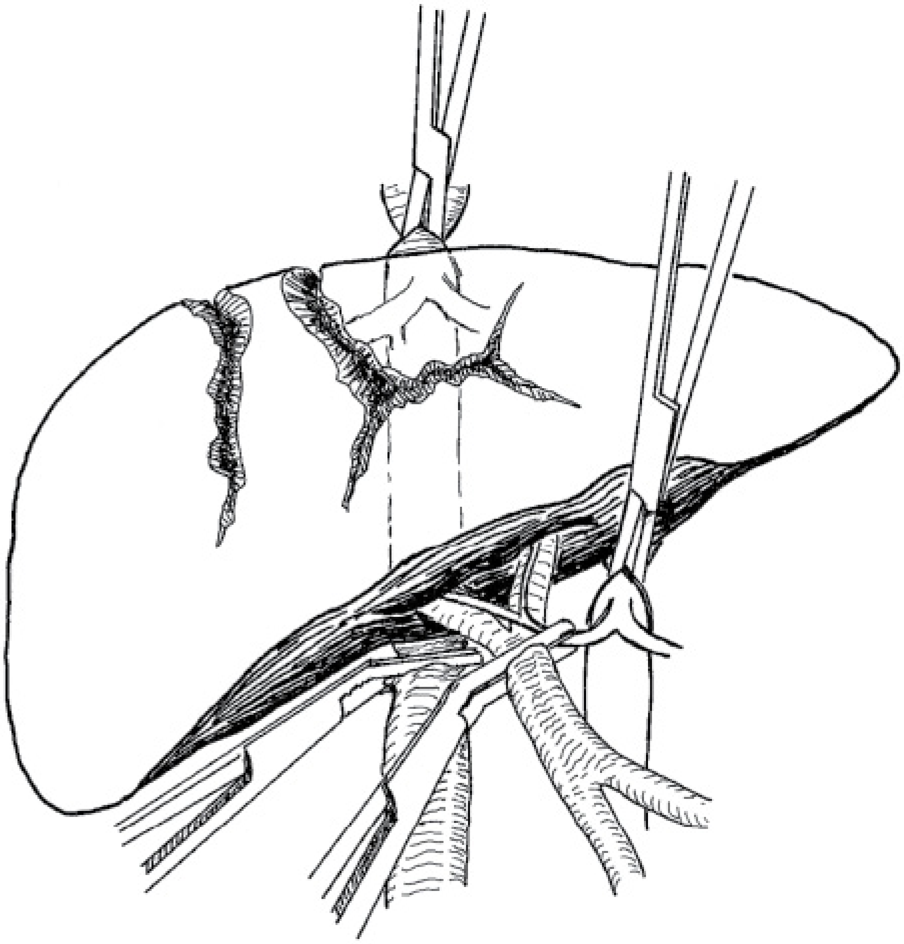

Blunt solid organ injury
Hemodynamically stable
Hemodynamically unstable
Transient response to resusciation
Persistent transfusion requirement
ANGIOGRAPHIC EMBOLIZATION
Successful
Persistent bleeding
Non-operative treatment
Laparotomy

84.5 Algorithm for selected use of angiographic embolization in patients with blunt solid organ injury (modified from M. Nance, CHOP, Philadelphia, PA, USA.)



84.6a,b

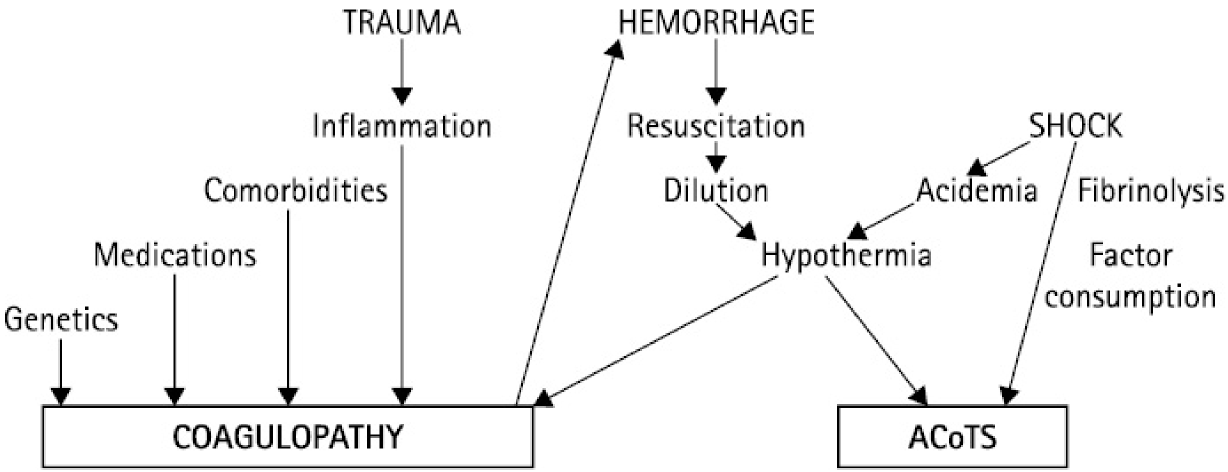
In young children with significant hepatic injury, the sternum can be divided rapidly to expose the suprahepatic or intrapericardial inferior vena cava, allowing for total hepatic vascular isolation with occlusion of the porta hepatis, suprahepatic and infrahepatic inferior vena cava, and supraceliac aorta (optional). Children will tolerate periods of vascular isolation as long as their blood volume is replenished. With this exposure, the liver and major perihepatic veins can be isolated and the bleeding controlled to permit direct suture repair or ligation of the offending vessel (**Figure 84.7**). Although the cumbersome and dangerous technique of atriocaval shunting has been largely abandoned, newer endovascular balloon catheters can be useful for temporary vascular occlusion to allow access to the juxtahepatic vena cava.



84.7

The early morbidity and mortality of severe hepatic injuries are related to the effects of massive blood loss and replacement with large volumes of cold blood products. The consequences of prolonged operations with massive blood product replacement include hypothermia, coagulopathy, and acidosis (**Figure 84.8**). Although the surgical team may keep pace with blood loss, life-threatening physiologic and metabolic consequences are inevitable, and many of these critically ill patients are unlikely to survive once their physiologic reserves have been exceeded. Maintenance of physiologic stability during the struggle for surgical control of severe bleeding is a formidable challenge even for the most experienced operative team, particularly when hypothermia, coagulopathy, and acidosis occur.

This triad creates a vicious cycle in which each derangement exacerbates the others, and the physiologic and metabolic consequences of the triad often preclude completion of the procedure. Lethal coagulopathy from dilution, hypothermia, and acidosis can occur rapidly. The infusion of activated recombinant factor VII in patients with massive hemorrhage has shown promising results.


TRAUMA
HEMORRHAGE
Inflammation
Resuscitation
SHOCK
Comorbidities
Dilution
Acidemia
Fibrinolysis
Medications
Hypothermia
Factor consumption
Genetics
COAGULOPATHY
ACoTS

84.8 Acute coagulopathy of trauma and shock.

Increased emphasis on physiologic and metabolic stability in emergency abdominal operations has led to the development of staged, multidisciplinary treatment plans including abbreviated laparotomy, perihepatic packing, temporary abdominal closure, angiographic embolization, and endoscopic biliary stenting. Abbreviated laparotomy with packing for hemostasis allowing resuscitation prior to planned reoperation is an alternative in unstable patients where further blood loss would be untenable. This "damage control" philosophy is a systematic, phased approach to the management of the exsanguinating trauma patient. The three phases of damage control are detailed in Table 84.2. Once patients are rewarmed, coagulation factors have been replaced, and oxygen delivery has been optimized, they can be returned to the operating room for pack removal and definitive repair of injuries.

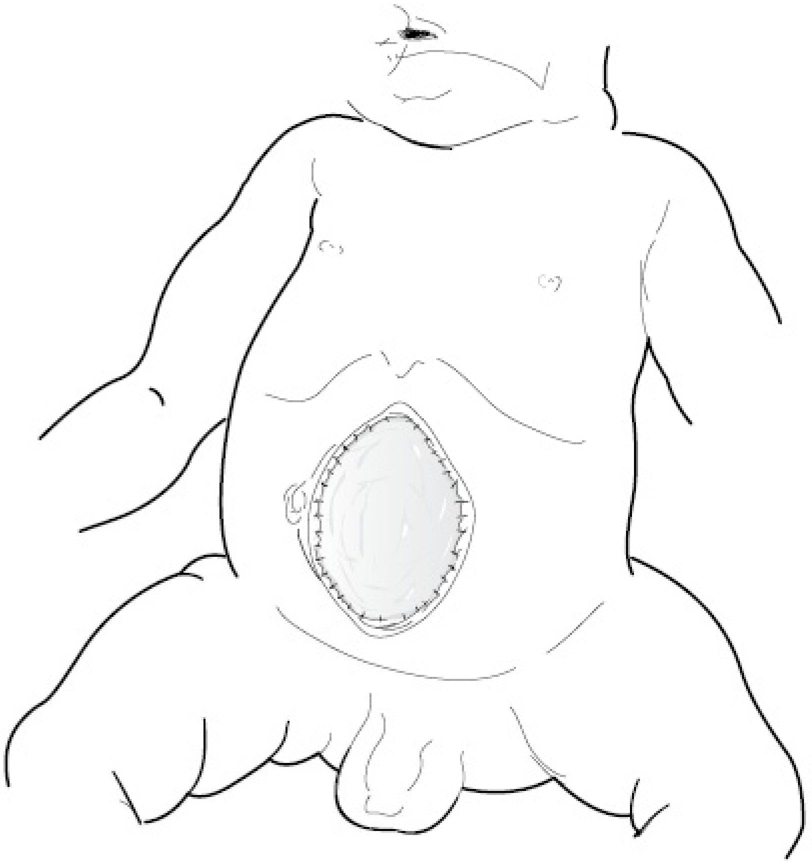
Table 84.2 **Damage control strategy in the exsanguinating trauma patient**

Phase	Action
Phase 1	Abbreviated laparotomy for exploration Control of hemorrhage and contamination Packing and temporary abdominal wall closure
Phase 2	Aggressive ICU resuscitation Core rewarming Optimize volume and oxygen delivery Correction of coagulopathy
Phase 3	Planned reoperation(s) for packing change Definitive repair of injuries Abdominal wall closure

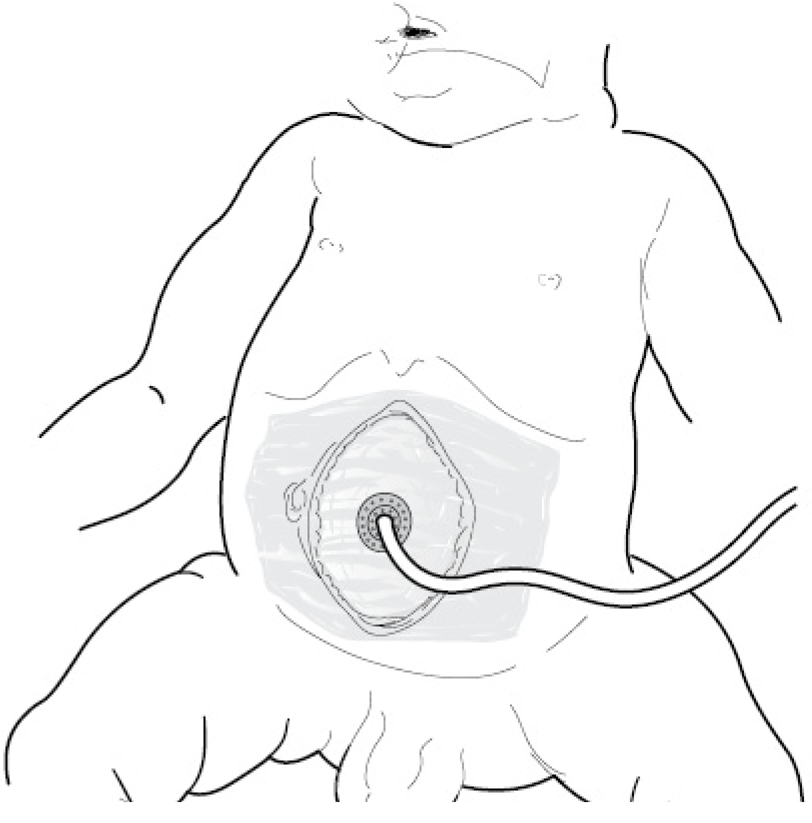
Abbreviation: ICU, intensive care unit.

While the success of abdominal packing is encouraging, it may contribute to significant morbidity, such as intra-abdominal sepsis, organ failure, and increased intra-abdominal pressure. It is essential to emphasize that the success of the abbreviated laparotomy and planned reoperation depends on an early decision to employ this strategy prior to irreversible shock. Abdominal packing, when employed as a desperate, last-ditch resort after prolonged attempts at hemostasis have failed, has been uniformly unsuccessful.

The obvious benefits of hemostasis provided by packing are also balanced against the potential deleterious effects of increased intra-abdominal pressure on ventilation, cardiac output, renal function, mesenteric circulation, and intracranial pressure. Timely alleviation of the secondary "abdominal compartment syndrome" may be a critical salvage maneuver for patients. Temporary abdominal wall closure at the time of packing can prevent the occurrence of this life-threatening syndrome. We recommend temporary abdominal wall expansion in all patients requiring packing until the hemostasis is obtained and visceral edema subsides. Many materials have been suggested for use in temporary patch abdominoplasty, including Silastic sheeting (**Figure 84.9a**), Gore-Tex® patches, IV bags, cystoscopy bags, ostomy appliances, and various mesh materials. The vacuum-pack technique, used successfully in adults, has been an outstanding addition in children (**Figure 84.9b**). Use of the vacuum-pack technique at the first trauma laparotomy may limit the early benefits of the open abdomen, which results in a lower volume reserve capacity.



(a)



84.9a,b

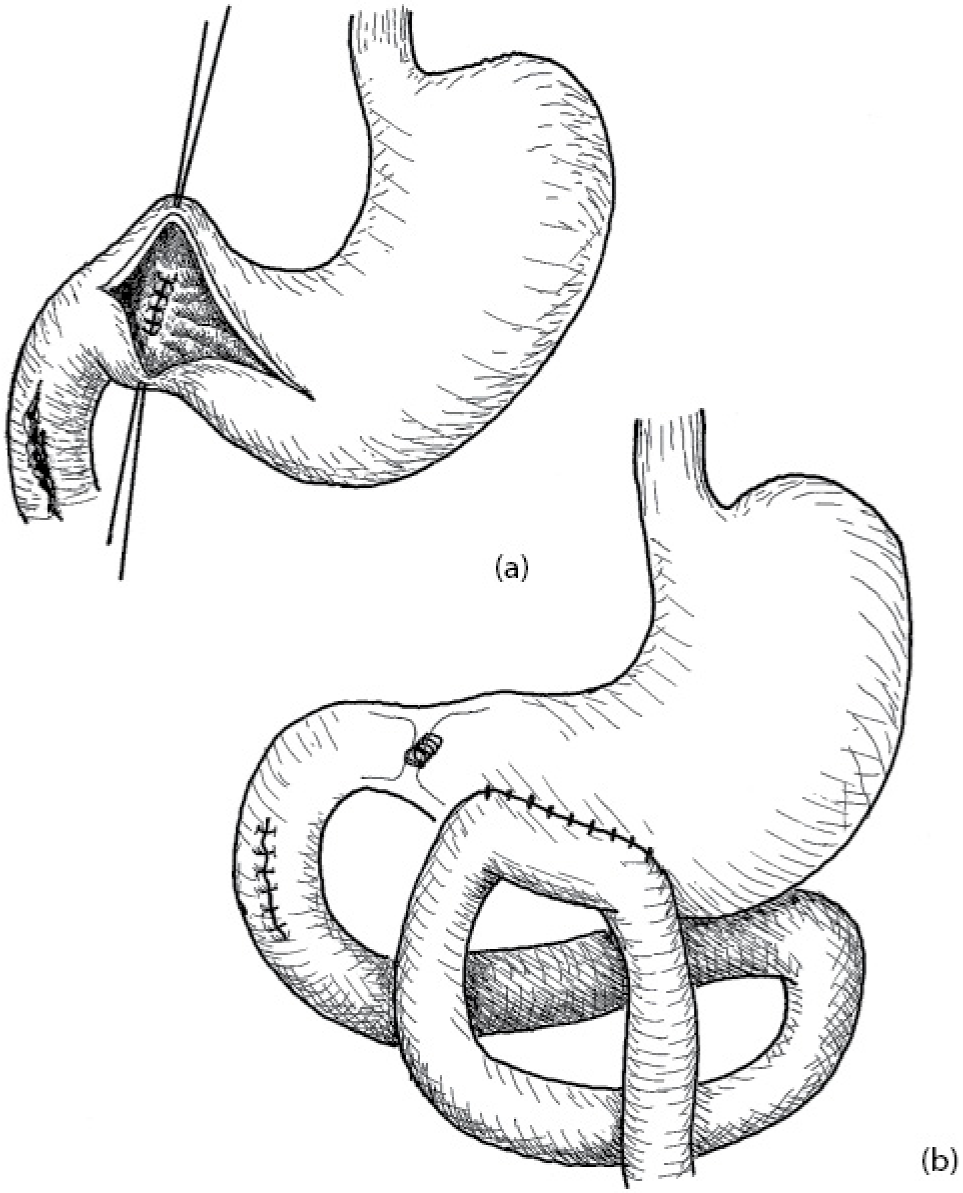
A staged operative strategy for unstable trauma patients represents advanced surgical care and requires sound judgment and technical expertise. Intra-abdominal packing for the control of exsanguinating hemorrhage is a life-saving maneuver in highly selected patients in whom coagulopathy, hypothermia, and acidosis render further surgical procedures unduly hazardous. Early identification of patients likely to benefit from abbreviated laparotomy techniques is crucial for success.

Duodenum and pancreas

Patients sustaining duodenal perforation are treated operatively in a variety of ways depending on the severity of the injury and the surgeon's preference:

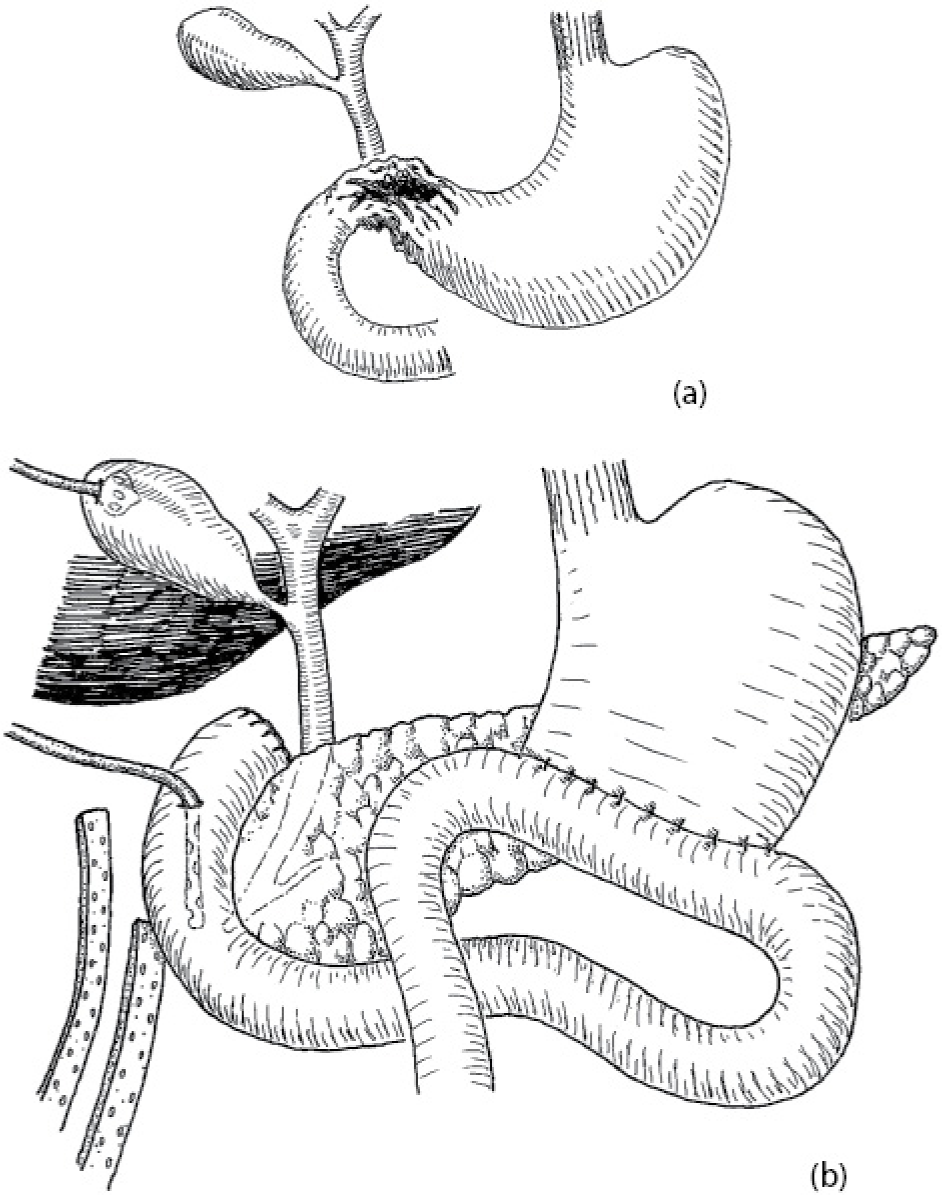
- Repair of the duodenum
- Diversion of the GI tract (pyloric exclusion or a duodenal diverticulization)
- Gastric decompression (gastric tube insertion or gastrojejunostomy)
- GI tract access for feeding (jejunostomy tube or gastrojejunal anastomosis)
- Decompression of the duodenum (duodenostomy tube)
- Biliary tube drainage
- Wide drainage of the repaired area (lateral duodenal drains)

The authors recommend primary closure of the duodenal perforation whenever possible (**Figure 84.10a**). Extensive lateral duodenal injury should be treated by primary duodenal repair and pyloric exclusion consisting of temporary closure of the pylorus with an absorbable suture and gastrojejunostomy (**Figure 84.10b**). Closed suction drainage of the repair is not depicted in this illustration. Feeding jejunostomy is often added to the procedure. When the duodenum is excluded, complete healing of the injury routinely occurs prior to the spontaneous reopening of the pyloric channel and spontaneous closure of the gastrojejunostomy.



84.10a,b

Duodenal diverticularization is an effective procedure for combined proximal duodenal and pancreatic injury. Resection and closure of the duodenal stump with decompressive tube duodenostomy, biliary drainage via tube cholecystectomy, gastrojejunostomy, and multiple closed suction drains are depicted in **Figure 84.11a and b**. A feeding jejunostomy should be strongly considered (not depicted). No matter what repair the surgeon selects, a summary of the literature demonstrates that protecting the duodenal closure (drain and exclusion) and a route for enteral feeds (gastrojejunostomy ± feeding jejunostomy) reduces morbidity and length of hospital stay. A pancreaticoduodenectomy (Whipple procedure) should rarely be required. Although occasionally reported in the literature, pancreaticoduodenectomy should be reserved for the most severe injuries to the duodenum and pancreas when the common blood supply is destroyed and any possibility of reconstruction is impossible.



84.11a,b

Injuries to the pancreas are slightly more common than duodenal injuries, with estimated ranges from 3% to 12% in children sustaining blunt abdominal trauma.

Reports from major pediatric trauma centers are clearly in conflict. Some favor and document the efficacy and safety of observational care for virtually all pancreatic traumas including ductal disruption, whereas others favor early distal pancreatectomy for transection to the left of the spine. It is clear that with simple transection of the pancreas at or to the left of the spine, spleen-sparing distal pancreatectomy can accomplish definitive care for this isolated injury with short hospitalization and acceptable morbidity. The procedure can be performed using minimally invasive techniques.

Treatment of children with blunt pancreatic injury has been debated for more than 25 years. Conclusions from past references were often biased by institutional preferences and expertise as well as the collection of cases with a wide range of injury severity. In particular, it was not often possible to decipher the presence or absence of major pancreatic ductal injury to allow for a relevant comparison.

Most would agree that minor (Grade I and II) pancreatic injuries heal without adverse sequelae without the need for surgery or invasive procedures. In addition, those with major pancreatic head disruption (Grade V) and concomitant duodenal injury require prompt surgery. Those patients with moderate to severe injuries (Grade III–IV) remain the topic of debate. Proponents of non-operative treatment cite a similar length of hospital stay when compared to operative cases and emphasize the adverse sequelae from distal pancreatectomy including pancreatic leak/fistulas, inadvertent splenectomy, and bowel obstruction. Those treated without surgery have a significantly higher rate of pancreatic pseudocysts requiring observation, percutaneous drainage, and occasional cyst-gastrostomy. Proponents of non-operative treatment acknowledge the higher pseudocyst rate yet indicate that the outcome after pseudocyst is often successful without the need for major resection. The largest series comparing outcomes in pediatric blunt pancreas injury included 14 pediatric trauma centers retrospectively reviewing 167 patients treated from 1995 to 2012. Patients with Grade II and Grade III injuries were selected for study. Patients with Grade III injuries made up the majority of the operative group. Outcomes for Grade III patients are in the **Table 84.3.**

Table 84.3 **Outcomes for Grade III pancreatic injuries**

Characteristic	Operative resection (n = 54)	Non-operative (n = 26)	p value
Pseudocyst (%)	0	44	<0.001
Need for repeat intervention (%)[a]	2	46	<0.001
Overall morbidity rate, %	33	61	0.05
Time to initial feeds (days)	4.4	12.7	0.01
Time to goal feeds (days)	9.4	26.1	0.01
Initial hospitalization (days)	11.9	15.0	n/s
Readmissions (%)	19	33	n/s
Mean total days in hospital	12.6	17.5	0.03
Mean time to complete resolution (days)	19.3	82.4	0.006

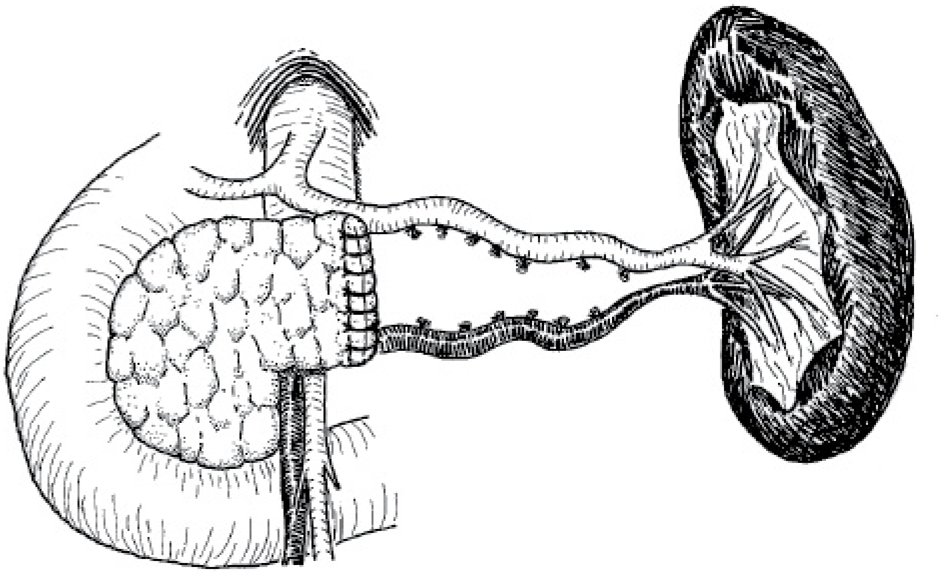
[a] Endoscopy or interventional radiology (exclusive of vascular access).

Despite the obvious limitations of a this multicenter, multi-year, retrospective review, Iqbal and colleagues have provided evidence supporting the conclusion that, in similar settings, one chooses to accept the higher rate of pseudocyst formation in non-operative treatment versus the morbidity associated with pancreatic resection after injury.

Laparoscopic distal pancreatectomy is often performed with four trocars. A hand-assist port can be useful in some settings, but use may be limited in younger children with less abdominal domain. Subcostal and perixiphoid trocar positions are modified according to the size of the child. Working ports should accept 5 mm instruments and at least one port should accept endosurgical stapling devices. After achieving pneumoperitoneum, the lesser sac is entered through the gastrocolic ligament and omentum. The pancreas is then explored through the lesser sac. If the spleen is to be preserved, the short gastric vessels are preserved. To gain further exposure of the pancreas, the short gastric vessels can be taken up to the level of the gastroesophageal junction, but splenectomy will then be required. The splenic flexure is then mobilized to expose the inferior edge of the tail of the pancreas. The pancreas is mobilized out of the retroperitoneum by incising the peritoneum from the inferior edge of the pancreas to the inferior pole of the spleen. The pancreatic tail can then be mobilized and retracted medially. This dissection allows the splenic artery and vein to be isolated and divided with a vascular stapler or between clips.

Two techniques allow for splenic preservation. The first technique spares the main splenic artery and vein. This requires division and coagulation of the pancreatic vascular branches from the splenic artery and vein. In contrast to elective pancreatic resections, the retroperitoneal inflammation (saponification) and hematoma secondary to pancreas injury will often make meticulous dissection of small pancreatic vascular branches from the splenic artery and vein impossible. It is possible to spare the spleen while ligating the splenic artery and vein (Warshaw procedure), as the spleen will be maintained by the preserved short gastric vessels. The inferior mesenteric vein may also need to be secured and divided during the mobilization of the distal pancreas.

The pancreas is then divided using a vascular stapler. It is often hazardous to obtain circumferential dissection of the pancreas proximal to the injury. Stapling the proximal margin may not be technically feasible. Suture oversewing, tissue glues, and large drains have been used to secure the proximal margin of pancreas (**Figure 84.12**). Endoscopic stenting of the ampulla of Vater may decrease fistula formation.



84.12

Kidney

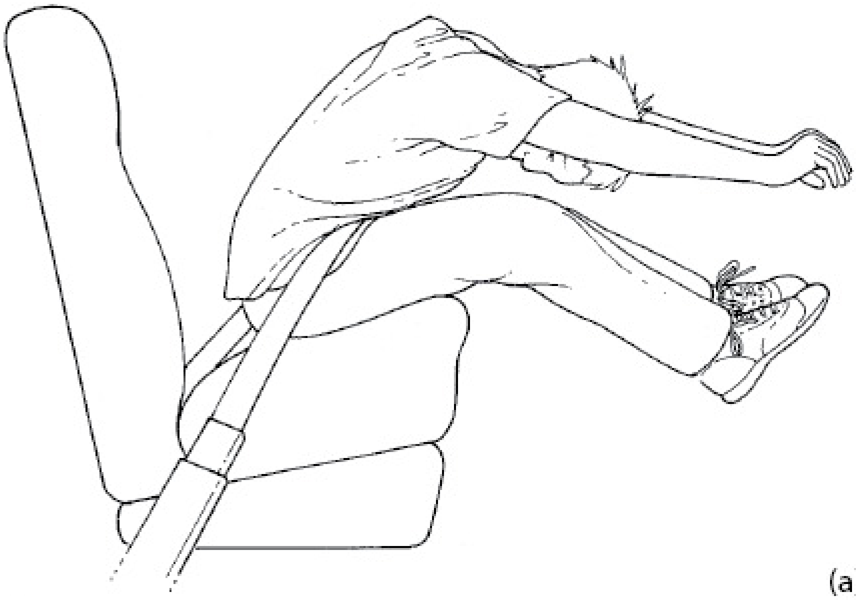
Renal injuries in children are often caused by high-energy impact associated with motor vehicle accidents or other serious abdominal trauma. Hematuria is present in 41–68% of children following blunt abdominal trauma. Most authors report direct correlation between the amount of hematuria and the severity of genitourinary injury, but renovascular injuries may result in no hematuria. Historically, congenital abnormalities in injured kidneys have been reported to vary from 1% to 21%. More accurate recent reviews have shown that incidence rates are 1–5%. Renal abnormalities, particularly hydronephrotic kidneys, may be first diagnosed after minor blunt abdominal trauma. Usually, these patients present with hematuria following blunt trauma.

It is imperative to acknowledge that major renal injuries, such as ureteropelvic junction (UPJ) disruption or segmental arterial thrombosis, may occur without the presence of hematuria or hypotension. Therefore, a high index of suspicion is necessary to diagnose these injuries. Nonvisualization of the injured kidney on IV pyelogram and failure to uptake contrast with a large associated perirenal hematoma on CT are hallmark findings for renal artery thrombosis. UPJ disruption is classically seen as perihilar extravasation of contrast with non-visualization of the distal ureter.

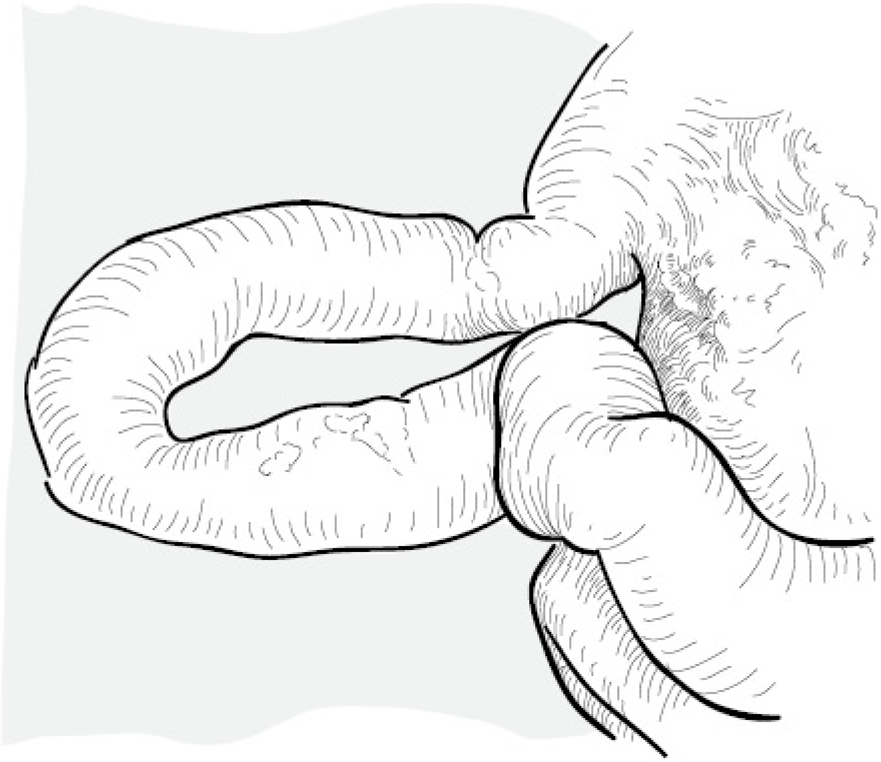
The majority of blunt renal injuries are treated without operation when uncontrolled hemorrhage or other indications for abdominal exploration are absent. This approach is safe and effective in 77–86% of children and most have excellent functional outcome without hypertension. Successful renal salvage at operation by partial nephrectomy or nephrorrhaphy depends on the severity of both the renal injury and associated injuries. Collecting system injuries should be repaired with absorbable sutures after evacuation of pelvic clots and debridement of devascularized parenchyma. Intravenous infusion of indigo carmine (a vital dye excreted in the urine) at operation may help identify sites of extravasation, and proximal control of the renal vessels prior to opening Gerota's fascia may facilitate retroperitoneal exploration. Early control of the vessels increases the rate of renal salvage. When proximal vascular control is performed before any renal exploration, nephrectomy is required in less than 12% of cases. When primary vascular control is not achieved and massive bleeding is encountered, the nephrectomy rate increases. Nephrectomy is recommended for major renal injuries in hemodynamically unstable patients with multiple injuries, and in those patients with avulsion injuries. Vascular repair can be attempted within 12 hours of injury and in the absence of multiple injuries.

The "seat-belt syndrome"

A decrease in motor vehicle-related fatalities has occurred in association with mandatory seat-belt legislation. Concurrently, a pattern of injuries caused by lap-belt restraints has emerged. This "seat-belt syndrome" includes abdominal wall contusion, injury to a hollow viscus, and vertebral fracture. The mechanism of injury is hyperflexion of the torso caused by deceleration forces with the lap-belt as a fulcrum (**Figure 84.13a**). Children are at particular risk for these injuries due to their higher center of gravity, thin abdominal wall, non-prominent iliac crest, and immature supporting structures of the vertebral bodies. Bowel injuries associated with lap-belt use can occur by several mechanisms: Compression against the vertebral column with a crush injury, shearing of the bowel and mesentery at fixed points in the retroperitoneum, and immediate closed-loop burst injuries on the antimesenteric border when fluid-filled loops are subjected to sudden increases in intraluminal pressure. The crush and shearing mechanisms may lead to progressive ischemia with delayed perforation or stricture (**Figure 84.13b**). In recent reports from Philadelphia, a database created by the State Farm Insurance Company was used to review 147 985 children who were passengers in motor vehicle crashes. In that series, 1967 children (1.33%) had abdominal bruising from seat-belt restraints. Although abdominal wall bruising was infrequent, those with this finding were 232 times more likely to have a significant intra-abdominal injury than those without a bruise. These data further revealed that one of nine children with an abdominal seat-belt sign had a significant intra-abdominal injury. Children with the “seat-belt sign” across their lower abdomen should be admitted for serial examinations even if the initial examination and diagnostic tests are normal (**Figure 84.13c**).



(a)



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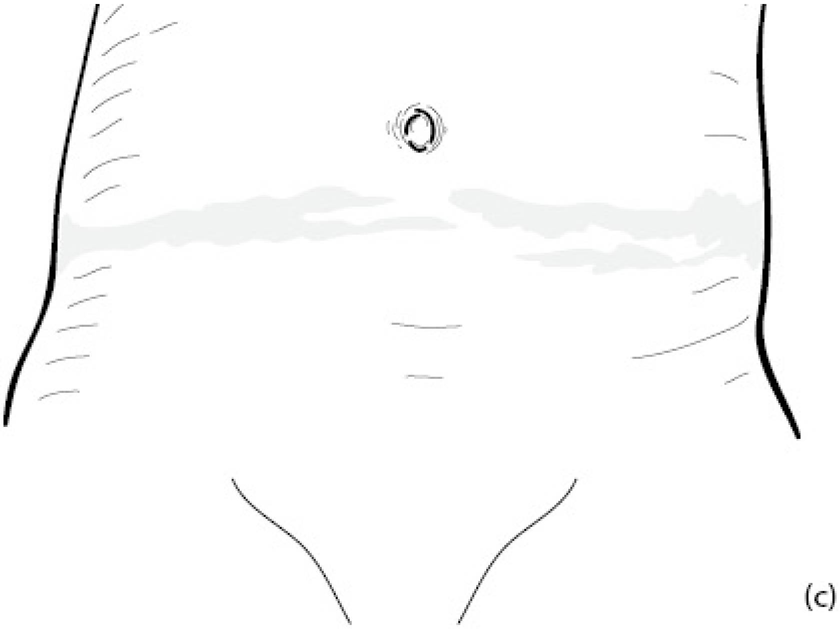


84.13a–c

CONCLUSION

The essential ingredient of pediatric trauma care is commitment to the special needs of injured children – personal, institutional, and community commitment. Recent advances in the delivery of trauma and critical care in children have resulted in improved outcome following major injuries. Incorporation of newer minimal access, endoscopy, and interventional radiology techniques is strongly urged. Although an increasing emphasis on non-operative treatment has occurred in the last two decades, the pediatric surgeon should remain the principal physician in the multidisciplinary care of these critically injured children.

The decision not to operate is always a surgical decision.

ACKNOWLEDGMENT

Original **Figures 84.2, 84.7, and 84.10–84.12,** are by Mark Mazziotti, MD.

FURTHER READING

Fenton SJ, Sandoval KN, Stevens AM, Scaife ER. The use of angiography in pediatric blunt abdominal trauma patients. *J Trauma Acute Care Surg* 2016; 81(2): 261–5.

Iqbal CW, Levy SM, Tsao K, et al. Laparoscopic versus open distal pancreatectomy in the management of traumatic pancreatic disruption. *J Laparoendosc Adv Surg Tech A* 2012; 22(6): 595–8.

Iqbal CW, St Peter SD, Tsao K, et al. Operative vs nonoperative management for blunt pancreatic transection in children: multi-institutional outcomes. *J Am Coll Surg* 2014; 218: 157–62.

LeeVan E, Zmora O, Cazzulino F, et al. Management of pediatric blunt renal trauma: A systematic review. *J Trauma* 2016; 80: 519–28.

Lutz N, Nance ML, Kallan MJ, et al. Incidence and clinical significance of abdominal wall bruising in restrained children involved in motor vehicle crashes. *J Pediatr Surg* 2004; 39: 972–5.

Naik-Mathuria BJ. Pancreatic Trauma Study Group. Proposed clinical pathway for nonoperative management of high-grade pediatric pancreatic injuries based on a multicenter analysis: A pediatric trauma society collaborative. *J Trauma Acute Care Surg* 2017; 83(4): 589–96.

Notrica DM, Eubanks JW 3rd, Tuggle DW, et al. Nonoperative management of blunt liver and spleen injury in children: Evaluation of the ATOMAC guideline using GRADE. *J Trauma Acute Care Surg* 2015; 79(4): 683–93.

Stylianos S. APSA Trauma Study Group. Prospective validation of evidence-based guidelines for resource utilization in children with isolated spleen or liver injury. *J Pediatr Surg* 2002; 37: 453–6.

PART IX

Special section

85

Vascular anomalies

EILEEN M. DUGGAN AND STEVEN J. FISHMAN

HISTORY

Vascular anomalies have been noted in history and folklore for centuries. Vascular birthmarks were often attributed to maternal impressions while the affected person was *in utero* and so were termed mother's marks (e.g. nevus maternus). This incorrect terminology has continued with generic terms such as "hemangioma" being applied to a wide variety of vascular lesions. Thus, misdiagnosis and misunderstanding of the natural history and treatment options for afflicted patients is common.[1]

PRINCIPLES AND JUSTIFICATION

The International Society for the Study of Vascular Anomalies (ISSVA) has accepted the terminology proposed by Mulliken in 1982 that classifies lesions broadly into vascular tumors and vascular malformations (**Table 85.1**).[2,3] Vascular tumors are proliferative lesions that range from benign to malignant. Vascular malformations consist of vessels that formed abnormally during development. They are classified by the type of vascular channel of which they are comprised, i.e. capillary (C), lymphatic (L), venous (V), or arterial (A). Malformations (M) can be combinations of more than one type (e.g. CLM) and they can be associated with non-vascular anomalies such as in Klippel–Trenaunay syndrome (CLVM + limb overgrowth), Parkes Weber syndrome (CAVM + limb overgrowth), CLOVES (*c*ongenital*l*ipomatous *o*vergrowth, *v*ascular malformations, epidermal *n*evi, and *s*keletal anomalies)[4] and Sturge–Weber syndrome (facial and leptomeningeal CM + eye anomalies ± bone and/or soft-tissue overgrowth).

Table 85.1 Abbreviated ISSVA classification for vascular anomalies

Vascular tumors			Vascular malformations	
Benign	**Locally aggressive**	**Malignant**	**Simple**	**Combined**
Infantile hemangioma Congenital hemangioma Tufted angioma Spindle-cell hemangioma Epithelioid hemangioma Pyogenic granuloma	Kaposiform hemangioendothelioma Retiform hemangioendothelioma PILA, Dabska tumor Composite hemangioendothelioma Kaposi sarcoma	Angiosarcoma Epithelioid hemangioendothelioma	Capillary malformation (CM) Lymphatic malformation (LM) Venous malformation (VM) Arteriovenous malformation (AVM) Arteriovenous fistula (AVF)	CVM, CLM LVM, CLVM CAVM CLAVM

Infantile hemangiomas are the most common vascular anomaly. They typically become apparent within the first week or two after birth, proliferate for approximately 10 months and then slowly involute. Most hemangiomas can be observed and allowed to involute spontaneously. Complicated or threatening hemangiomas can be treated with propranolol or other medications to accelerate involution.

Vascular malformations do not involute spontaneously and often require treatment. Due to the broad pathology of these lesions, evaluation and treatment by a multidisciplinary team is often required. Some lesions can be observed or require only non-invasive therapies such as compression or medical therapy. One of the most common medications in current use for vascular malformations is sirolimus, an mTOR inhibitor often used for complex lymphatic malformations or lesions with significant lymphatic components. Sclerotherapy is used for venous or lymphatic malformations as primary treatment or to shrink lesions prior to resection. Operative treatment of vascular malformations can be done in lesions not amenable to these other therapies or are more often done in combination with these other therapies. Though the range of operations done for these lesions is wide, we will focus our discussion of technique to operations performed by general pediatric surgeons that are not often used for other conditions.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

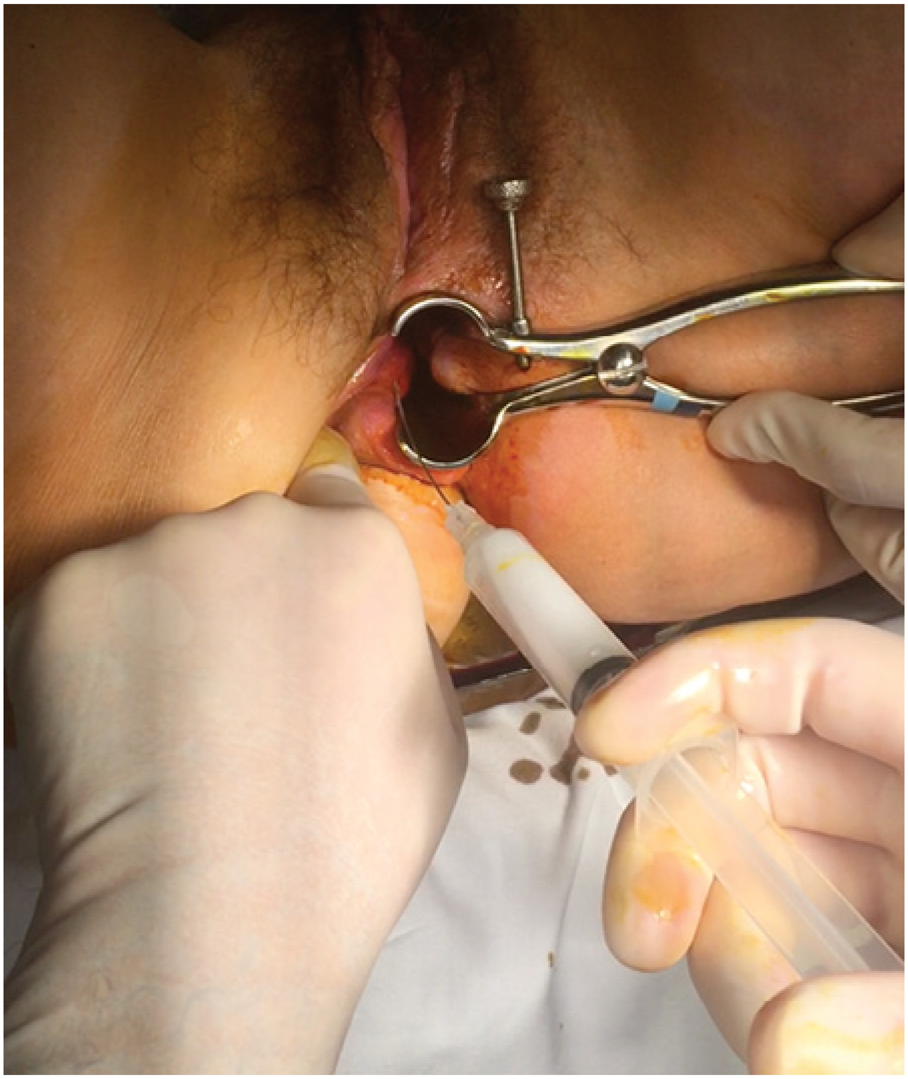
In patients with certain anomalies that are more high-risk (e.g. multifocal or extensive VMs, LVMs, CLVMs), we recommend more extensive preoperative hematologic evaluation. These patients are more likely to have chronic consumptive coagulopathies. While the coagulopathy does not normally cause serious problems at baseline, it can worsen with surgical resection or other major procedures and lead to serious systemic bleeding and thrombotic complications. Therefore, preoperative plasma d-dimer, fibrinogen, platelets, PT, and PTT should be drawn. Concerning numbers include a d-dimer more than 20 times the upper limit, fibrinogen levels <100 mg/dL, elevated PT or PTT, and thrombocytopenia. In cases of lab abnormalities, replacement therapy with platelet or cryoprecipitate should be given perioperatively to keep fibrinogen greater than 100 mg/dL and platelet counts over 100 000/μL. The administration of low-molecular weight heparin may bring fibrinogen to normal levels without the need for cryoprecipitate.

In addition to coagulation abnormalities, patients with CLVM and other overgrowth disorders such as CLOVES syndrome are at higher risk for pulmonary emboli.[5] Many of these patients have ectatic embryonic or central veins that contribute to the risk already inherent to their prothrombotic state. Preoperative vein closure with embolization, sclerotherapy, or endovenous ablation may therefore be needed for non-essential ectatic veins prior to major procedures. Prophylactic anticoagulation or caval filtration should also be considered. We recommend consulting a hematologist perioperatively for discussion of thrombotic prophylaxis and other hematologic issues in these complicated patients.

RECTAL SCLEROTHERAPY

Patients with symptomatic rectal venous malformations may be treated with sclerotherapy.

Patients are placed in lithotomy position and the buttocks are elevated with padding. Anoscopy is first done to visualize the most distal extent of the disease. If it extends proximally, rigid proctoscopy is then done, looking for the point where the abnormal red vascular tissue transitions to normal salmon-pink mucosa. The anoscope is then used to expose two quadrants of the rectum. Sclerosant is drawn up into a 10 mL syringe with a 22-gauge long spinal needle; we use a 1 : 13% sodium tetradecyl sulfate (STS) to saline solution mixed with air until foamy. The spinal needle pierces into the mucosa right above the dentate line and is advanced through the extent of the malformation, staying in the submucosal plane (**Figure 85.1**). The sclerosant is then injected while the needle is pulled out – the malformation should become pale from the underlying sclerosant. After both accessible quadrants are injected, the anoscope is turned 90 degrees and this process is repeated in the remaining two quadrants. A piece of compressed gel foam is then placed in the rectum for hemostasis. This will be expelled with the patient’s first bowel movement.

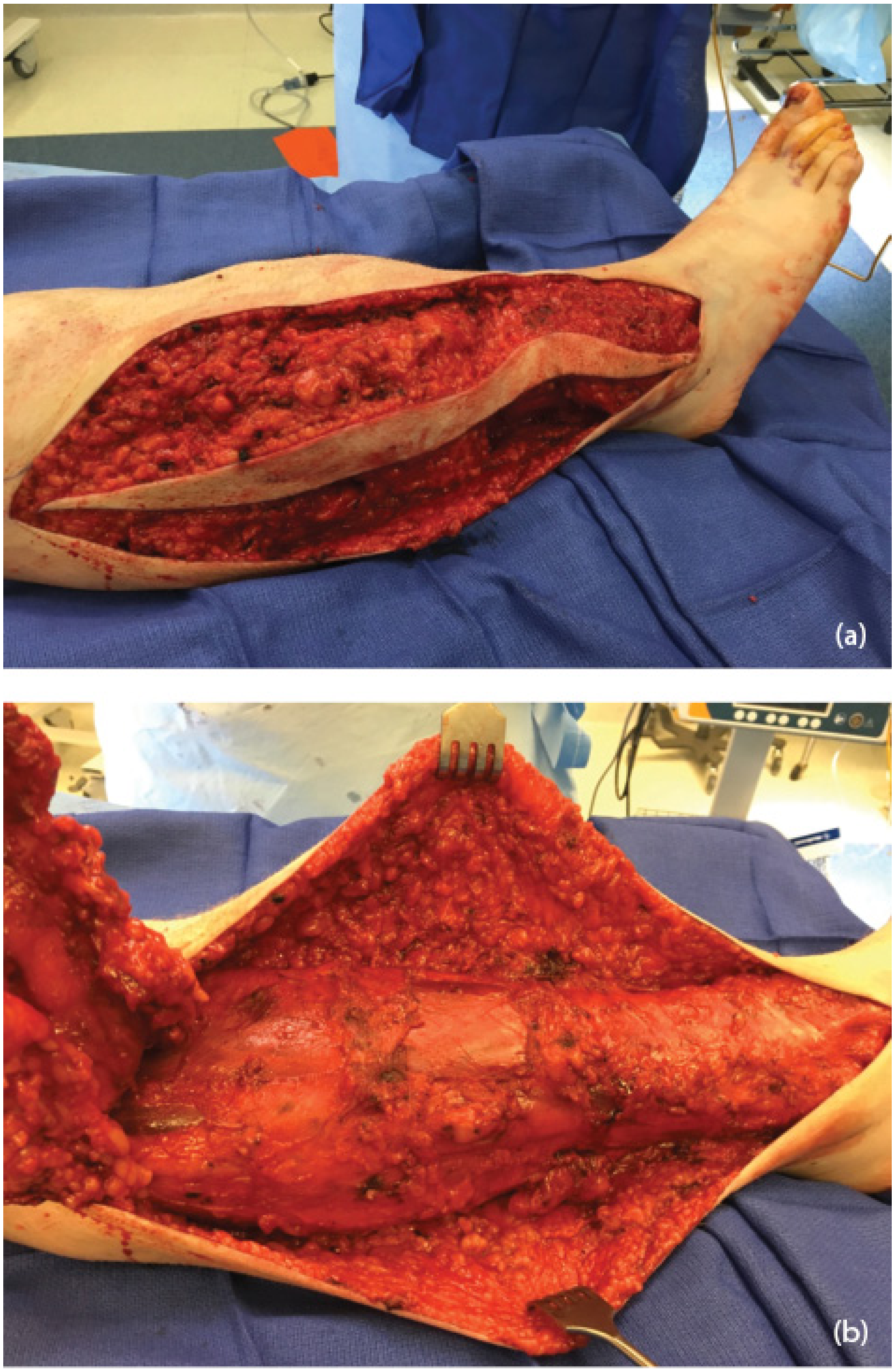


85.1 Rectal sclerotherapy.

VASCULAR MALFORMATION DEBULKING

Patients with excessive extremity, truncal, buttocks, perineal, or genital bulk secondary to their vascular anomalies and associated overgrowth may benefit from debulking procedures. Benefits of this can include improvement in mobility, decrease in soft-tissue infections, and psychologic improvement secondary to improved esthetics and ability to fit into clothes or shoes. Debulking is most effective when the excess tissue is mostly located superficial to muscle fascia.

If working on the extremity, a tourniquet should be considered for lesions with significant venous malformation. A lenticular skin excision is made when excess skin is present, incorporating the most affected skin if possible (**Figure 85.2a**). If buttock debulking is being done, the incision should be made in the infragluteal fold. Skin flaps are then made in all directions. The affected tissue between the dermis and the underlying fascia is removed, taking care to leave just enough tissue for a well-vascularized flap (**Figure 85.2b**). In genital debulking it is best to avoid being overly aggressive, with a goal to achieve symmetry rather than to remove all malformation. After hemostasis is assured, the flaps are closed over drains.[6]

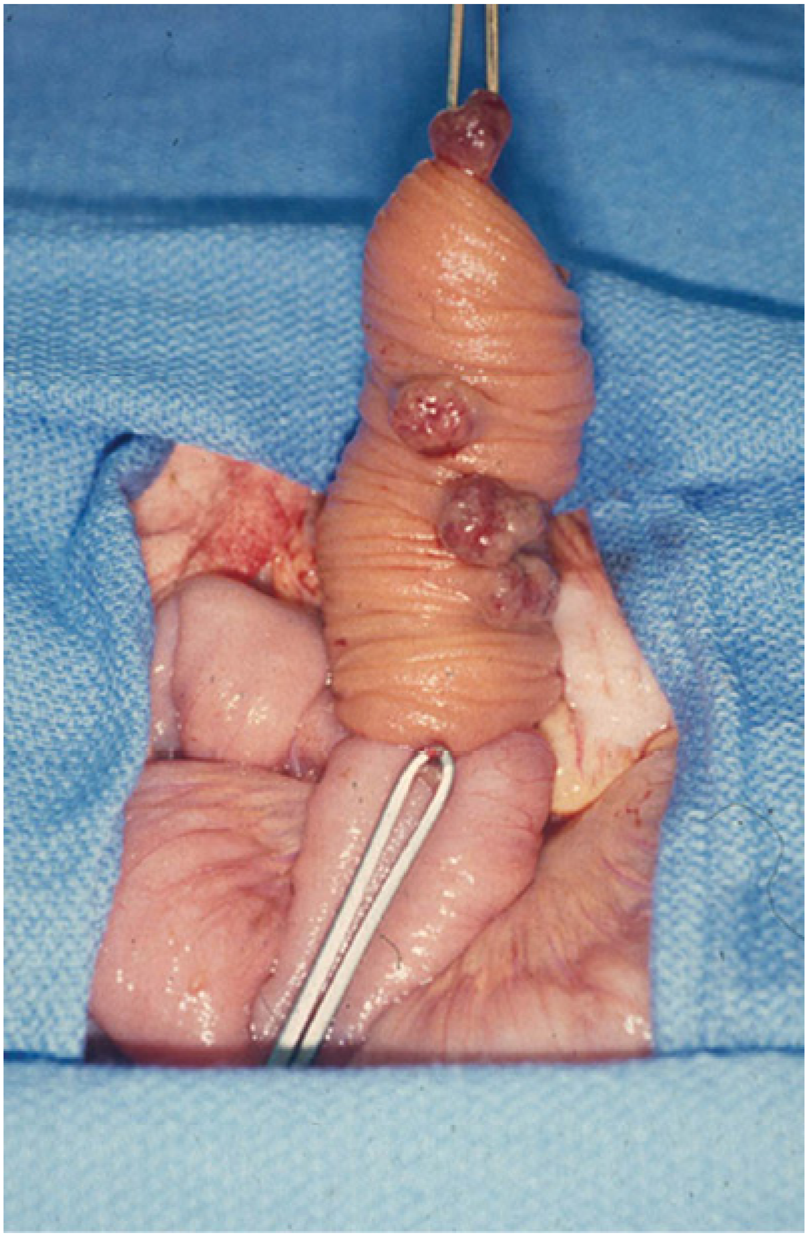


85.2a,b Suprafascial debulking of lower extremity with (a) lenticular skin excision with flap development and (b) affected tissue removal.

EXCISION OF MULTIFOCAL GASTROINTESTINAL LESIONS IN BLUE RUBBER BLEB NEVUS SYNDROME

Patients with blue rubber bleb nevus syndrome (BRBNS) have multifocal venous malformations located mostly in the skin, soft tissues, and gastrointestinal (GI) tract. In some patients, these lesions can cause GI bleeding severe enough to require regular blood transfusions. In addition, the lesions can occasionally cause pain or obstruction due to intussusception. In these cases, we have often done lengthy operations with a goal of removing all detectable GI lesions while resecting no or as little intestinal length as possible.[7] For patients with diffuse lesions that would preclude significant operative excision or for younger patients in whom we would like to avoid an operation until later in their life, sirolimus therapy may be a good alternative.[8]

After preoperative bowel preparation, the patient is placed in supine position on the operating room table. A midline laparotomy is done and any adhesions are freed so that the bowel is mobile. Intraoperative upper and lower endoscopy is then carried out with manual assistance to telescope the bowel over the endoscope in order to examine all mucosal surfaces of the entire GI tract. All lesions are marked with a suture externally for later identification. Lesions are then excised with a combination of mucosal resections, full-thickness wedge resections, and limited bowel resections for densely involved areas. We attempt to access as many lesions as possible through a single enterotomy by manual prolapse of the proximal and distal bowel through that defect (**Figure 85.3**). Esophageal or rectal lesions difficult to access via the peritoneal cavity can be treated by endoscopic sclerotherapy, band ligation, or transanal excision. A nasogastric tube is left in place and the abdomen is closed.



85.3 Prolapsed intestinal mucosa showing multifocal venous malformations.

POSTOPERATIVE CARE

Subcutaneous drains should be left in place after most extremity, torso, and genital debulking procedures. We expect prolonged drainage from these wounds, sometimes for several months, especially in the case of large amounts of lymphatic tissue. Drains should be removed when the drainage is less than 10–15 mL per day.

The nasogastric tube is left in place for patients with BRBNS until bowel function returns.

COMPLICATIONS

Rectal sclerotherapy can cause a feeling of fullness in about 90% of patients, but 10% of patients will have exquisite perianal pain similar to a thrombosed hemorrhoid. Rarely, sclerosed areas can undergo mucosal ulceration afterwards and significant bleeding. This can often be managed with rectal packing. Sclerotherapy usually needs to be repeated an average of every 1–2 years for good control. In cases of failure of sclerotherapy to control bleeding or desire to avoid repeated procedures, some patients may progress to needing a proctectomy and endorectal pull-through.

Wounds from debulking procedures often heal poorly. The possibility of wound breakdown or leakage after the operation should be discussed with the family preoperatively. Wound breakdown is cared for with standard wound care methods.

Malformation regrowth is possible after debulking procedures as there is likely microscopic vascular malformation left over even if all visible affected tissue is removed. In addition, lymphatic vesicles can often form at the site of the resection scar.

After resection of GI vascular malformations, an ileus lasting a week or more is not uncommon due to extensive bowel manipulation. Intestinal leaks and wound infections are rare. Appearance of lesions several years after bowel resection has been seen in several patients, more commonly in those undergoing an operation pre-adolescence. If bleeding becomes a problem several years after the operation, repeated endoscopy should be done and another eradication procedure may need to be performed.

OUTCOME

While there are very few vascular anomalies that can be cured, we aim to improve patients' quality of life with interdisciplinary therapy. Quality of life may include esthetic or functional improvement, decreased pain, or decreased bleeding/leaking. Evidence of improvement may include increased hemoglobin/hematocrit, decreased transfusion requirements, decreased incidence of infections, or improved validated quality-of-life measures.

REFERENCES

1. Mulliken JB, Burrows PE, Fishman SJ. *Mulliken and Young's Vascular Anomalies*. Oxford: Oxford University Press, 2013.
2. International Society for the Study of Vascular Anomalies (ISSVA). Classification. Available at: https://www.issva.org/classification [Accessed 26 June 2020.]
3. Wassef M, Blei F, Adams D, et al. Vascular anomalies classification: Recommendations from the International Society for the Study of Vascular Anomalies. *Pediatrics* 2015; 136(1): e203–14.
4. Alomari AI. Characterization of a distinct syndrome that associates complex truncal overgrowth, vascular, and acral anomalies: A descriptive study of 18 cases of CLOVES syndrome. *Clin Dysmorphol* 2009; 18(1): 1–7.
5. Alomari AI, Burrows PE, Lee EY, et al. CLOVES syndrome with thoracic and central phlebectasia: Increased risk of pulmonary embolism. *J Thorac Cardiovasc Surg* 2010; 140(2): 459–63.
6. Kulungowski AM, Fishman SJ. Management of combined vascular malformations. *Clin Plast Surg* 2011; 38(1): 107–20.
7. Fishman SJ, Smithers CJ, Folkman, et al. Blue rubber bleb nevus syndrome: Surgical eradication of gastrointestinal bleeding. *Ann Surg* 2005; 241(3): 523–8.
8. Salloum R, Fox CE, Alvarez-Allende CR, et al. Response of blue rubber bleb nevus syndrome to sirolimus treatment. *Pediatr Blood Cancer* 2016; 63(11): 1911–14.

86

Fetal surgery

EMILY A. PARTRIDGE AND ALAN W. FLAKE

HISTORY

The first description of open maternal–fetal surgery for the correction of anatomic anomalies was reported by Michael R. Harrison and colleagues at the University of California, San Francisco, in 1983, with the successful creation of bilateral ureterostomies in a fetus with congenital bilateral hydronephrosis due to urinary tract obstruction. Since then, great progress has been made in the ability to diagnose fetal abnormalities, predict outcomes, and perform prenatal surgical intervention when appropriate. The concept of the fetus as a patient has become a standard of care and the ethical framework for maternal fetal intervention is now well developed. While application of open fetal surgery has remained limited to a relatively small number of highly selected fetuses and is practiced in only a few centers, the development of this field has accelerated technological progress in prenatal diagnosis and intervention, led to improved understanding of the pathophysiology and natural history of candidate disorders, allowed comprehensive counseling of parents in centers with focused expertise in fetal anomalies, and driven the evolution of less invasive therapeutic approaches.

PRINCIPLES AND JUSTIFICATION

Maternal–fetal surgery is a specialty born of clinical necessity; a congenital defect alters normal development and causes irreversible organ damage before birth, leading to prenatal or neonatal death. Observations of fetuses with congenital anomalies and neonates with irreversible end-organ damage led to the compelling rationale that correction *in utero* might arrest progression of disease or even reverse the pathophysiology and restore normal development. Improvements in fetal imaging and serial clinical observation have allowed better definition of fetal pathophysiology, and better prediction of which fetuses might benefit from prenatal intervention. Because of the potential maternal risk, maternal–fetal surgery has historically been reserved for fetal disorders deemed to have a high probability of causing fetal or neonatal death. However, the success of the Management of Myelomeningocele Study (MOMS) in reducing the neurologic morbidity associated with myelomeningocele (MMC) has expanded the potential application of maternal–fetal surgery to non-lethal conditions.

The term "maternal–fetal surgery" includes a procedural spectrum ranging from ultrasound (US)-guided shunt placement to image-guided fetoscopic procedures, to the more invasive open fetal surgery and *ex utero* intrapartum treatment (EXIT) procedures. Minimally invasive procedures have become feasible due to the development of fetoscopic equipment in recent years, and most can be performed entirely percutaneously with local anesthesia and less maternal morbidity than open surgery. The EXIT procedure, originally developed for use in fetuses undergoing tracheal occlusion for congenital diaphragmatic hernia (CDH), is now used in a variety of disorders in which difficulty obtaining an airway or achieving adequate ventilation is anticipated (**Table 86.1**). Despite the progress in minimally invasive surgery, open fetal surgery continues to be required for correction of most fetal structural anomalies.

Table 86.1 Indications for EXIT

Indication	Condition
Airway obstruction	Cervical teratoma Lymphangioma/lymphatic malformation Congenital high airway obstruction syndrome (CHAOS) Tracheal/laryngeal atresia Laryngeal web Severe micrognathia/hypoplastic craniofacial syndrome
Chest masses	Congenital cystic adenomatoid malformation (CCAM) Bronchopulmonary sequestration Mediastinal/pericardial teratoma
Circulatory support	Congenital diaphragmatic hernia (CDH) with associated cardiac malformation Congenital heart disease (hypoplastic left heart syndrome) Sacrococcygeal teratoma (SCT) – debulking procedure

There have been two recent randomized controlled multicenter clinical trials supporting fetal intervention. The MOMS trial demonstrated improvement in neurologic morbidity of MMC with prenatal closure, and the European trial demonstrated efficacy of fetoscopic laser separation versus amnioreduction in twin-to-twin transfusion syndrome (TTTS). Further randomized trials are clearly needed prior to broad application of controversial procedures, such as fetoscopic balloon tracheal occlusion for CDH, however they would be impractical and potentially unethical in rare anomalies requiring fetal intervention such as congenital cystic adenomatoid malformation (CCAM), and sacrococcygeal teratoma (SCT), and in circumstances of clear rationale and demonstrated efficacy such as the EXIT procedure for obstructive airway lesions.

KEY PREOPERATIVE INVESTIGATIONS AND PREPARATION

The prerequisites for consideration of open fetal surgery include:

- An accurate prenatal diagnosis and exclusion of associated anomalies
- The presence of a correctable lesion that, if left untreated, will lead to fetal demise or severe irreversible organ damage before birth

- Well-defined natural history of the disorder allowing selection of fetuses that will benefit from prenatal intervention
- Technical feasibility of fetal surgery with an acceptable risk-to-benefit ratio for both mother and fetus

Contraindications to fetal surgery include complex chromosomal or associated anatomic abnormalities in the fetus, maternal risk factors including incompetent cervix, placentomegaly, maternal mirror syndrome, morbid obesity, any serious comorbid conditions, including prohibitive psychiatric or psychosocial disorders, drug or alcohol abuse, and heavy smoking.

Patients suspected of carrying a fetus with a major anomaly should be referred to a fetal treatment center for comprehensive multidisciplinary evaluation. Evaluation should include detailed ultrasonographic characterization, fetal echocardiography, and often ultrafast magnetic resonance imaging (MRI). Fetal US was first described in the late 1960s, and remains the primary imaging modality for prenatal screening and diagnosis with its proven utility, relatively low cost, and widespread availability. Further, ultrasonography is advantageous due to its multiplanar capability, Doppler flow depictions, high spatial resolution, and real-time assessment. Limitations of ultrasonography include a relatively small field of view, beam attenuation by maternal adipose tissue, poor image quality in oligohydramnios, poor acoustic access to the fetal head when it lies deeper in the pelvis, and limited visualization of the posterior fossa due to calvarial calcification later in gestation.

When fetal intervention is a consideration, diagnostic certainty supported by accurate and specific details is paramount to the plan of care, and MRI is a useful adjunct and may be required for specific anomalies. The advantages of MRI include a larger field of view, superior soft-tissue contrast, more precise volumetric measurements, and greater accuracy in demonstrating intracranial abnormalities. However, long-term safety after exposure to high-field MRI has not been demonstrated, and there are concerns for effects on embryogenesis, chromosomal structure, and fetal development. As a precaution, MRI is not recommended during the first trimester. Furthermore, gadolinium contrast crosses the placenta and has been found in animal studies to be associated with growth retardation, so gadolinium contrast is not recommended in pregnant patients.

Prospective parents should undergo detailed non-directive counseling based on the results of this workup. All available options for the pregnancy should be presented, along with the risks and benefits of each. If open fetal surgery is offered, the mother must be counseled regarding the need for Cesarean section in this and all future pregnancies, and care must be taken to "allow" eligible families to choose non-surgical management, including termination or palliative care. A fetal karyotype or rapid fluorescent *in situ* hybridization (FISH) analysis for major chromosomal abnormalities should be performed. In general, significant associated anomalies, chromosomal abnormalities, or maternal risk factors such as heavy smoking, obesity, serious comorbid conditions, placentomegaly, and the maternal mirror syndrome are considered to be contraindications to open maternal–fetal surgery.

OPERATION: OPEN MATERNAL–FETAL SURGERY

Anesthesia

Patients should be admitted prior to any planned open procedure for monitoring and initiation of tocolysis. Because any invasive fetal procedure involves the risk of preterm labor, separation/rupture of the membranes, and chorioamnionitis, a 50 mg indomethacin suppository should be given 6 hours preoperatively for tocolysis and IV cefazolin should be given prior to incision. Tocolysis should be continued postoperatively with an IV 6 g loading dose of magnesium sulfate, followed by a continuous infusion at 2–4 g/hour for 18–24 hours. Magnesium levels should be closely monitored, and patients should be assessed for any clinical signs of magnesium toxicity during this period. Indomethacin suppositories should be given every 6 hours for the 48 hours following the procedure. After this time, patients may be converted to an oral nifedipine regimen of 10–20 mg every 6 hours, continued until delivery.

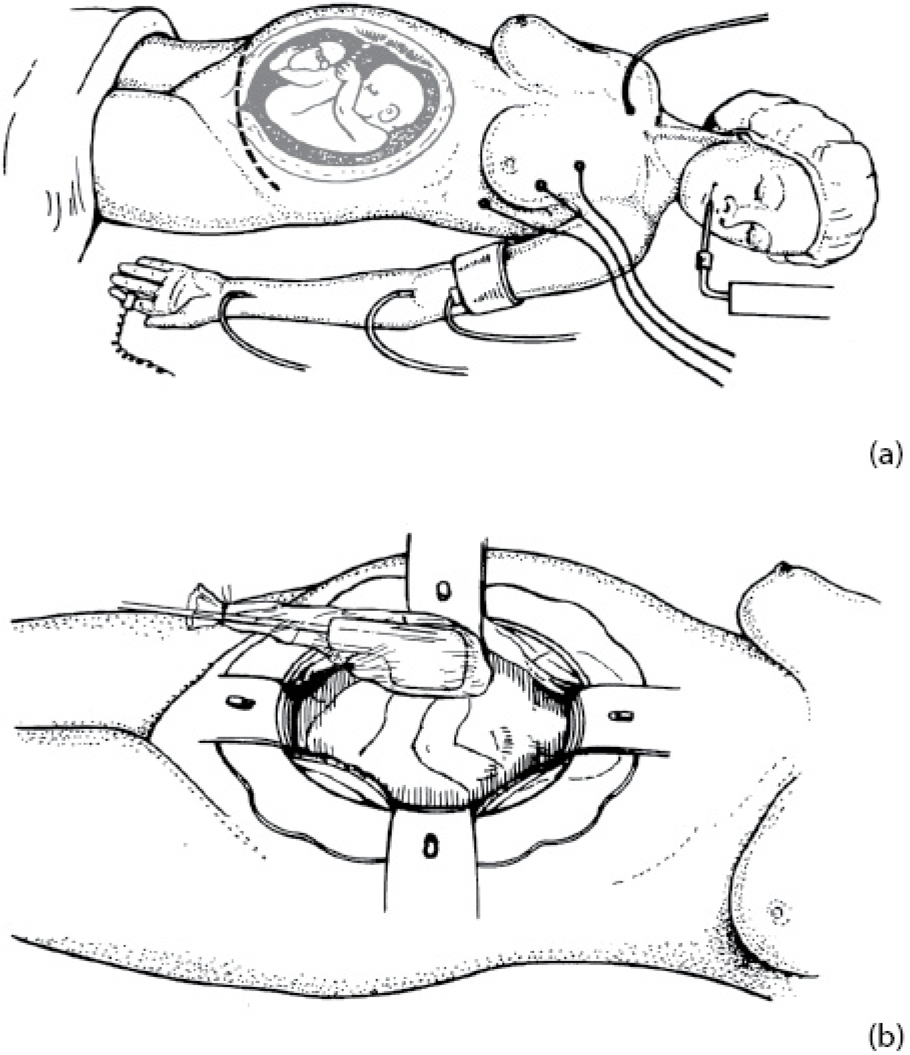
In order to ensure adequate uterine relaxation, an epidural catheter should be placed preoperatively and deep inhalational general anesthesia should be induced. Maternal monitoring should include a radial arterial catheter and blood pressure cuff, bladder catheter, electrocardiogram leads, and pulse oximetry, while access should include multiple large-bore IV catheters. Sequential compression devices should be employed to prevent deep venous thromboses.

Positioning should be supine with left lateral tilt, to minimize aortocaval compression from the gravid uterus. The patient is positioned supine with left lateral tilt obtained. A type and screen is sufficient for most minimally invasive procedures, but open procedures require available cross-matched blood for the mother, and warm type O-negative blood for the fetus, which can be cross-matched with the maternal sample to avoid reaction with maternal antibodies that cross the placenta.

Because the mother and fetus have separate, though codependent, anesthetic concerns, both an obstetric and a pediatric anesthesiologist are necessary. A sonographer/echocardiographer should be part of the surgery team, and a high-resolution US machine with color Doppler should be used to identify fetal and placental anatomy and to assess for potential hazards, such as velamentous cord insertion. During the procedure, continuous echocardiography should be used in combination with pulse oximetry to monitor fetal heart rate, cardiac function, and volume status.

Maternal incision

The initial incision is generally a low transverse abdominal incision (**Figure 86.1a**). Precise mapping of the placenta is paramount because placental position determines the position of hysterotomy placement.



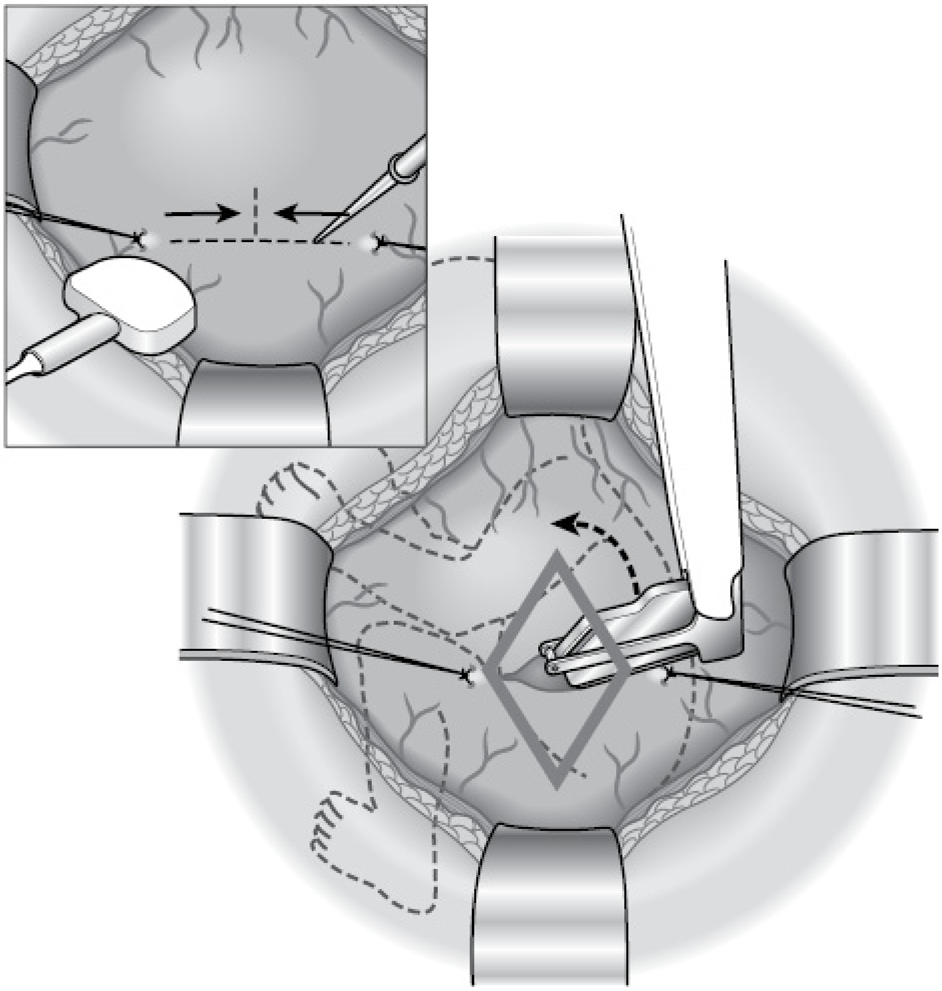
86.1a,b

For any placental position, skin flaps should be raised to allow exposure through a vertical midline fascial incision extending from the umbilicus to the symphysis pubis (**Figure 86.1b**). With a posterior placenta, the uterus will remain in the abdomen during the procedure. In cases with an anterior placenta, the uterus must be tilted out of the abdomen, and care is required to avoid compression of the lateral uterine vessels by the fascia and anterior compression from the abdominal wall or assistants while a posterior hysterotomy is performed. Once the uterus is lifted out of the abdomen, towels can be placed behind it to support its position and the patient can be returned to a flat orientation from the left lateral tilt position.

Hysterotomy

Prior to hysterotomy, the uterus is palpated and examined to determine whether sufficient relaxation has been achieved. It should be soft to palpation and there should be no visible striations on its surface. Transuterine US confirms fetal and placental position, and electrocautery is used to map the placental margins on the surface of the uterus. The hysterotomy site should avoid uterine vasculature and be at least 6 cm from the placental edge. The lower segment of the uterus is avoided due to increased risk of amniotic fluid leak, chorioamnionitis, and preterm labor.

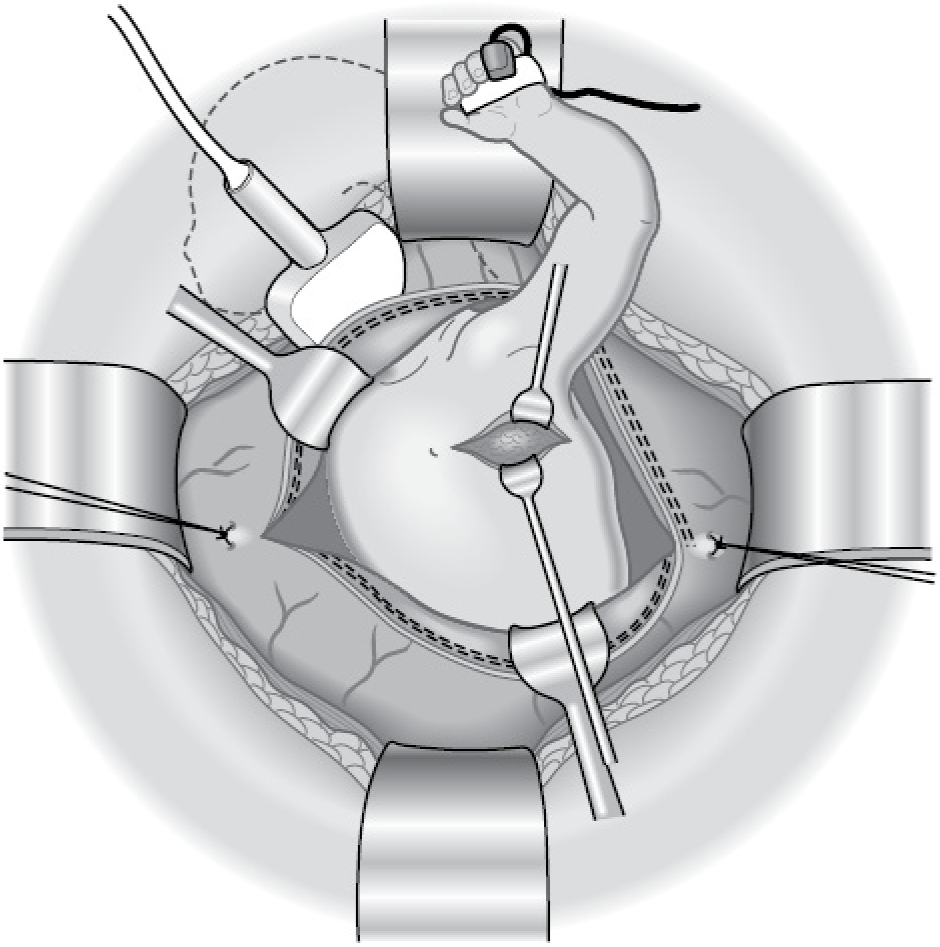
Once a site is chosen, opposing 0 PDS traction sutures are placed through the uterine wall and fetal membranes under US guidance. Using electrocautery, a 2 cm incision is made between the sutures. Angled vascular clamps are placed in a parallel configuration to compress the uterine wall in the planned position of the staple line prior to placing the stapler. A uterine stapler is then placed through the opening and fired once in either direction to compress the myometrium and minimize blood loss during hysterotomy, while maintaining membrane integrity for closure (**Figure 86.2**). Absorbable staples are used to avoid subsequent fertility issues. Using a rapid infuser, warmed lactated Ringer's solution is infused into the amniotic space via a catheter to maintain amniotic fluid volumes and fetal temperature, while preventing cord compression.



86.2

Fetal exposure

Following hysterotomy, the fetus is positioned within the uterus to expose the body part of interest (**Figure 86.3**). Continuous fetal echocardiography should be used to monitor cardiac function and ventricular rate. Depending upon the procedure and exposure, a fetal IV catheter is placed for infusion of fluids, blood, or medications, and intramuscular fetal anesthesia, and paralysis is provided with fentanyl and vecuronium. A pulse oximeter should be wrapped around the fetal hand or foot and covered with aluminum foil and Tegaderm™. Operative repair should proceed in standard fashion, taking care to keep the fetus warm, buoyant, and as submerged as possible.



86.3

Procedure: CCAM debulking

While the administration of maternal corticosteroid therapy has significantly decreased the development of high-output cardiac failure in the setting of congenital lung lesions, maternal–fetal surgery is required in a subset of pregnancies in which an inadequate response to betamethasone results in persistent hydrops fetalis. The arm and hand on the affected side are exposed, and the fetus is rotated to expose the chest wall, leaving the head and remainder of the body within the amniotic sac. Once IV access is obtained and a pulse oximeter is attached, the fetus is treated with atropine and volume loaded to counter reflexive bradycardia and cardiovascular collapse, which are often seen with acute decompression of the chest when the tumor is exposed. Electrocautery is used to create a large posterolateral thoracotomy at the sixth intercostal space. The lobe containing the CCAM is exteriorized, the attachments to surrounding lung tissue are divided, and the lobar pulmonary artery is ligated prior to ligation of the vein and bronchus in order to avoid lobar congestion. The bronchus is ligated next, followed by the pulmonary vein. Great care should be taken on exteriorization of lower-lobe tumors not to tear the delicate inferior pulmonary vein. The thoracotomy is then closed followed by uterine closure. Delivery following maternal–fetal surgery should be planned as late as possible to avoid complications associated with prematurity.

Procedure: SCT debulking

The evolution of high-output physiology and secondary hydrops in a fetus with SCT is nearly always associated with fetal demise and supports the rationale for performing open fetal surgery. Fetuses with large Type I tumors exhibiting clear evidence of early hydrops related to tumor flow prior to 27 weeks gestation are candidates for open fetal surgery with debulking of the tumor. Fetal intervention aims to prevent progression of vascular steal phenomenon and high-output physiology. If hydrops or placentomegaly should develop after 27 weeks, early delivery with debulking is recommended. This allows stabilization of the critically ill newborn in the neonatal intensive care unit prior to definitive resection.

The hysterotomy site is chosen to allow exteriorization of the tumor and the caudal end of the fetus. However, the umbilical cord is at risk for compression against the rim of the hysterotomy in this position, and the fetus should be continuously monitored for signs of cord compression. Care must be taken to keep the remainder of the fetus within the amniotic sac; inadvertent delivery of the whole fetus can lead to uterine contraction, inability to place the fetus back in the amniotic sac, and preterm labor.

Once the tumor is exteriorized, a Hegar dilator should be placed in the rectum to delineate anatomy, and the skin around the anorectal sphincter is incised, keeping an adequate margin of tissue between the incision and the sphincter complex. Fetal skin around the base of the tumor is then incised, controlling the large subcutaneous veins with suture ligatures as necessary. A tourniquet is applied around the base of the tumor where the skin has been incised to restrict blood flow to the tumor. A hand-held Harmonic scalpel™ (Ethicon Inc., Cincinnati, OH, USA) or other sealing device is then used to divide the tumor at its base. As the tourniquet is released, any bleeding vessels are ligated with suture ligatures. Any intrapelvic component of the tumor should be left as well as the coccyx, to be excised at the time of definitive resection postnatally. Once the tumor bed is hemostatic, the skin can be closed over the tumor bed, the fetus can be returned to the amniotic cavity, and the hysterotomy can be closed.

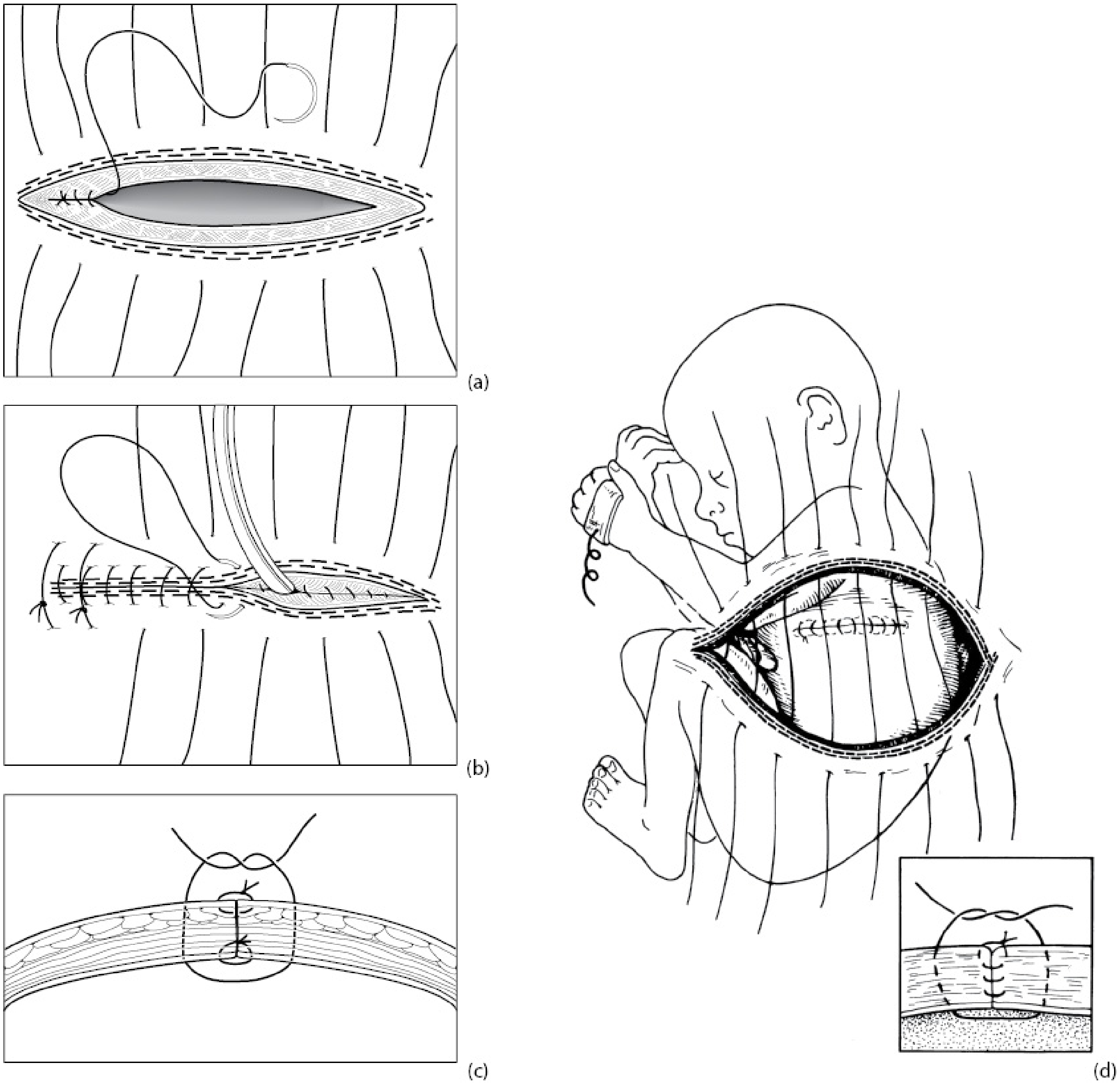
Procedure: Fetal MMC repair

Fetal MMC repair is offered to patients based on the inclusion criteria established in the MOMS trial, including singleton pregnancy with an MMC at level T1 through S1, Arnold–Chiari II malformation, gestational age 19^0 to 25^6 weeks, and normal karyotype without coexisting severe anomalies.

The operative procedure begins with a low transverse laparotomy, followed by creation of a hysterotomy as described in "Hysterotomy" previously. The fetus is positioned to expose the MMC or myeloschisis lesion. Continuous intraoperative fetal echocardiographic monitoring is critical. Fetal anesthesia is provided by the maternal inhalational anesthetic, and a narcotic dose and paralysis is delivered intramuscularly to the fetus. For MMC, the cystic membrane of the MMC is excised, the placode returned to the spinal canal, and the distal end of the placode/cord untethered. For myeloschisis the cord is untethered. In both lesions myofascial, dural-lined flaps are raised on each side of the defect. The flaps are reflected over the defect and closed with a running suture and then the skin, creating a thick, watertight closure. If the skin cannot be closed primarily, an acellular dermal graft is used to assist with the closure.

Uterine closure

The uterine closure must have adequate strength to prevent rupture and be watertight to prevent amniotic fluid leaks (**Figure 86.4a–d**). A two-layer closure should be performed, using double-armed full thickness 0 PDS stay sutures approximately 2 cm apart and 2 cm back from the staple line and a running 2/0 PDS suture through myometrium and membranes. Prior to completion of the running layer, approximately 400 mL of warmed lactated Ringer's solution containing 500 mg of oxacillin should be instilled into the amniotic cavity with US confirmation of adequate fluid volumes. An omental flap buttresses the uterine closure, and the maternal laparotomy should be closed in layers. A transparent Tegaderm™ dressing allows continued fetal monitoring postoperatively.



86.4a–d

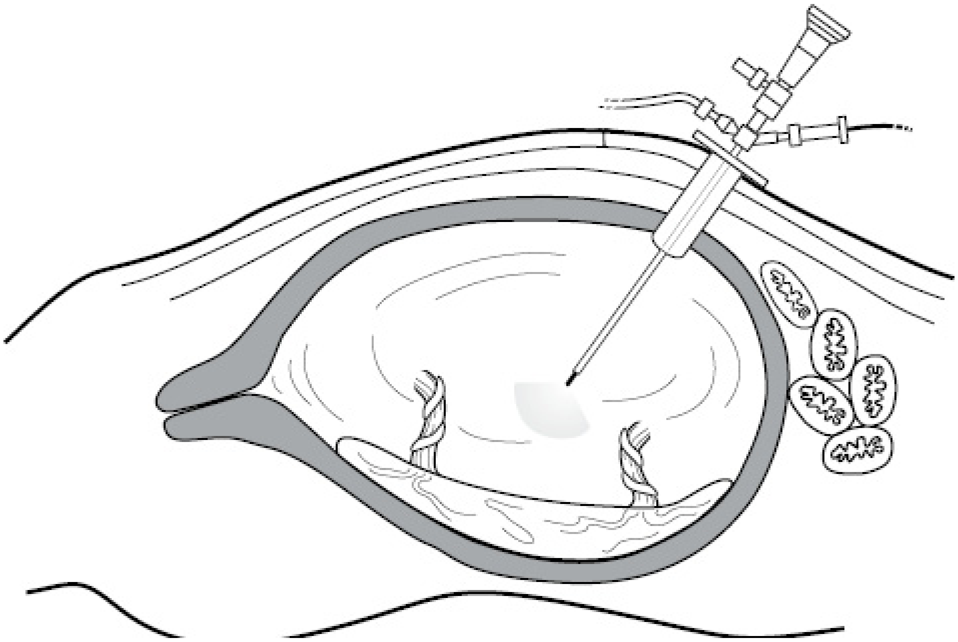
OPERATION: FETOSCOPY

When first developed in the 1970s, fetoscopy functioned as a diagnostic tool, but it has since become a tool for minimally invasive fetal interventions with the development of more advanced camera equipment and endoscopic devices. Fetoscopy has similar complications to open fetal surgery, including bleeding, preterm premature rupture of membranes, preterm labor with delivery <32 weeks, chorioamniotic separation, and chorioamnionitis. The risk of fetoscopic complications correlates with the size of the fetoscope, the number of trocar sites, and the length of the procedure. For simple procedures, the risk of preterm labor is lower than open fetal surgery. The procedure can generally be performed percutaneously under local or regional anesthesia. However, any procedure that would potentially cause fetal pain should include intramuscular injection of an opioid and a paralytic agent for fetal anesthesia.

Minimally invasive approaches may offer lower risk of preterm labor than open fetal surgery, potentially changing the risk–benefit ratio for fetal intervention. Currently, fetoscopic approaches are being investigated for several indications, although the most commonly performed procedure is fetoscopic photocoagulation of anomalous placental anastomoses in TTTS.

Positioning

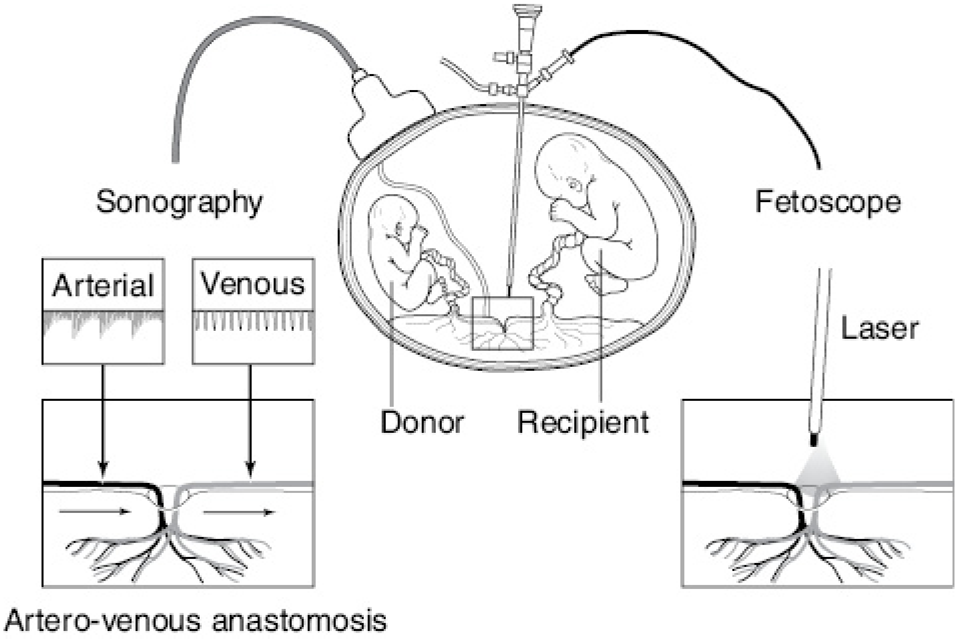
The patient is placed in low lithotomy position and intraoperative US is used to define the position of the placenta and fetus(es) (**Figure 86.5**).



86.5

Procedure

Laser fibers can be inserted through the side port of a fetoscopic operating sheath, allowing the use of a single trocar in TTTS (**Figure 86.6**). Under US guidance, unpaired vessels are coagulated. A stab incision is made for US-guided placement of an external trocar, usually ranging in diameter from 2 mm to 4 mm. For complex fetal interventions, an irrigating fetoscope can be used to exchange turbid amniotic fluid. The introduction of more than one trocar increases the risks of preterm labor and membrane disruption and leak. The majority of currently performed fetoscopic procedures are performed through a single trocar and extensively utilize image guidance. The risk of amniotic fluid leak is low with trocar sizes of 3 mm or less, and closure of the amniotic sac or fascia is not required. While more complex procedures may one day be possible using multiple trocars or single port technology, the capabilities of fetoscopy currently remain limited.



86.6

The current mainstay for treatment of TTTS is fetoscopic selective laser photocoagulation (SLPC), targeting the anastomoses that contribute to the imbalance of flow, performed between 18 and 26 weeks' gestation.

Historically, amnioreduction was the primary treatment modality for TTTS, but this is now rarely applied as primary therapy unless TTTS develops outside the gestational age at which SLPC is safe.

SLPC is performed percutaneously, most often under local anesthesia. A 2–3 mm fetoscope is inserted, with or without a trocar, under US guidance. The placental vasculature is mapped using direct visualization as well as Doppler US to identify anastomoses, and to define the placental equator. All anastomoses between the two placental circulations are targeted for ablation with a 30–50 W diode laser (**Figure 86.6**). Amnioreduction may also be performed at the end of the procedure if necessary to reduce intrauterine pressure. Because of the incidence of both early and late complications of SLPC, close follow-up is important for all patients.

OPERATION: EXIT PROCEDURE

The EXIT procedure was originally developed to allow removal of tracheal clips and establishment of an airway at the time of delivery after prenatal tracheal occlusion had been performed in severe cases of CDH. However, the indications for EXIT have expanded to include any case in which difficulty in obtaining an airway is anticipated, including CHAOS, giant anterior neck masses including cervical teratomas, pharyngeal tumors, mediastinal tumors, and hypoplastic craniofacial syndrome, or cases in which a thoracic mass will require resection in order to ventilate the patient, as in giant thoracic masses with dramatic mediastinal shift (large solid CPAMs).

Anesthesia

The main goal of the procedure is to maintain uteroplacental blood flow, providing gas exchange for the fetus while establishing an airway, resecting a neck or chest mass, or cannulating for extracorporeal membrane oxygenation (ECMO). This requires complete uterine relaxation and maintenance of uterine volume to avoid placental abruption and interruption of blood flow. Unlike a standard Cesarean section, the procedure is performed under a deep inhalational anesthetic in order to promote uterine relaxation. Fetal anesthesia is supplemented with intramuscular fentanyl and vecuronium for more complex procedures requiring operative dissection and/or fetal immobilization.

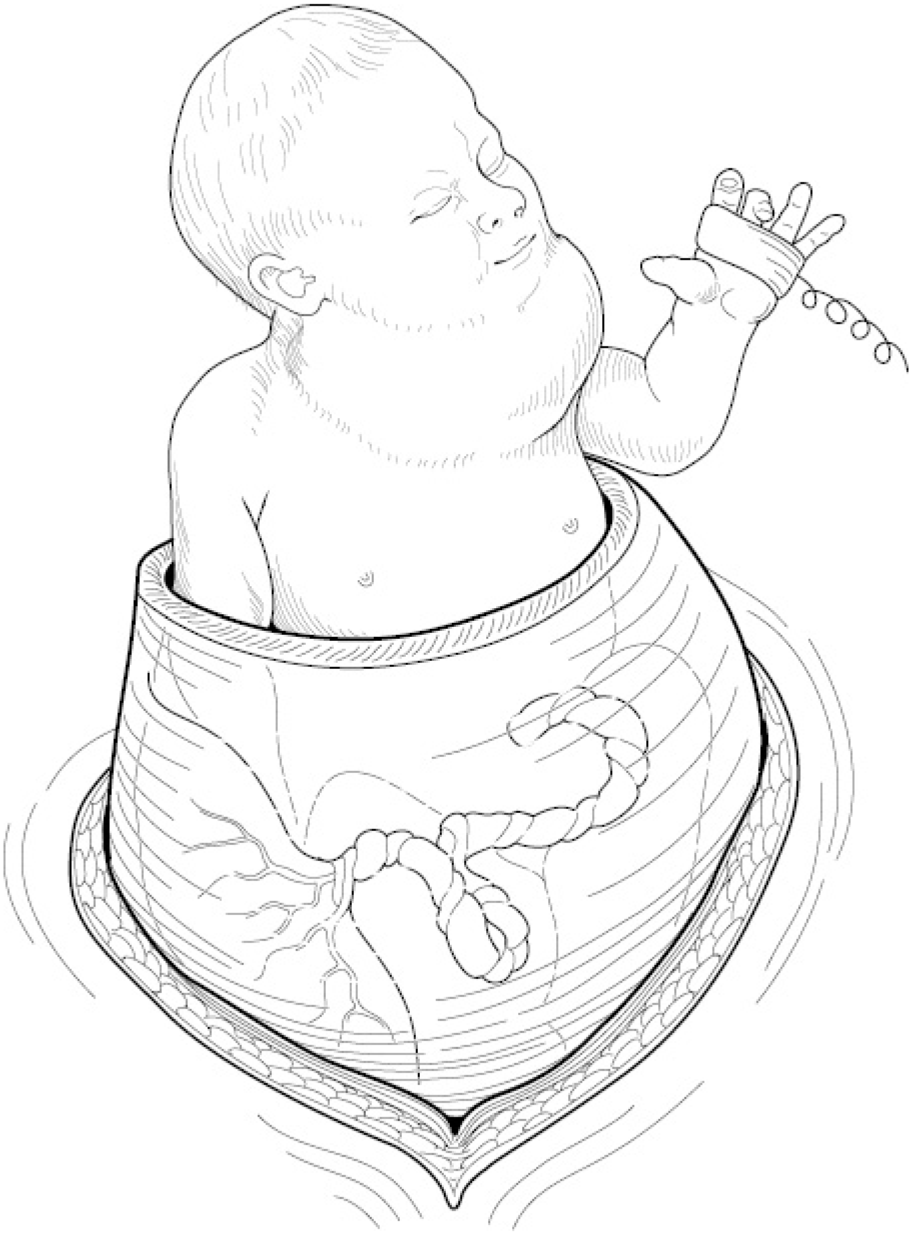
Similar to open maternal–fetal surgery, the patient is positioned supine, with left lateral tilt to maximize blood flow to the uterus and placenta as well as venous return. In addition to inhalational, intramuscular narcotic and paralytic agents are given to prevent fetal discomfort and movement.

During the procedure, uteroplacental blood flow must be maintained despite the vasodilatory effects of general anesthesia. Maternal blood pressure should be maintained within 10% of normal, and continuous fetal monitoring should assess for myocardial depression. As the fetus is delivered at the end of the procedure, tocolytics are not routinely required, but may be used synergistically with inhaled anesthetic for uterine relaxation. Prior to clamping the cord, measures should be taken to prevent excessive bleeding due to uterine atony, including discontinuation of inhaled anesthetic, administration of oxytocin, and uterine massage. Methergine and carboprost may also help to recover uterine tone in refractory cases.

A successful EXIT procedure relies on adequate uterine relaxation to maintain uteroplacental blood flow and prevent placental separation. However, this relaxation can lead to maternal postoperative bleeding. Additional maternal risks of EXIT include uterine rupture or scar dehiscence in a subsequent pregnancy, and wound infection. The goal is to have the fetus off placental support within 60 minutes, although procedures up to 2.5 hours have been reported. Risks to the fetus during EXIT include hypoxic brain injury, bradycardia, hemorrhage, and death secondary to cord compression, loss of myometrial relaxation, and placental abruption, all of which cause inadequate uteroplacental gas exchange.

Procedure

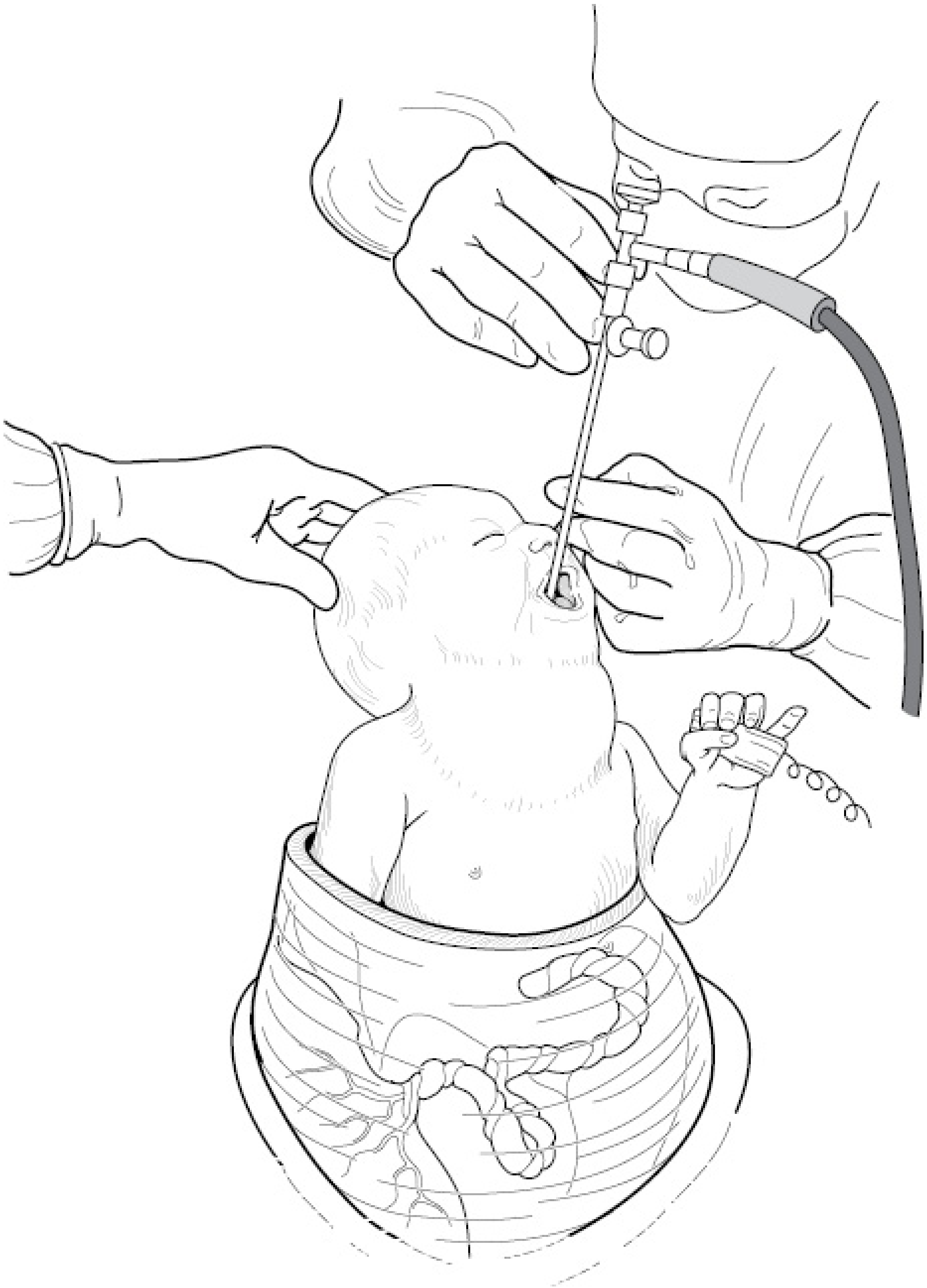
The maternal incision is carried out in a similar fashion to that performed in open maternal–fetal surgery. US is used to map the placental edges, and the uterus is opened with the same absorbable stapler used in open fetal surgery to prevent blood loss and control the membranes. In cases with severe polyhydramnios or large cystic masses, amnioreduction or cyst aspiration may be required before hysterotomy is performed. A peripheral IV line may be placed on the fetus, a pulse oximeter is applied to the exposed fetal hand (**Figure 86.7**), and continuous fetal transthoracic echocardiography is utilized to assess fetal cardiac function and volume status. Warmed Lactated Ringer's solution is infused to maintain uterine volume and fetal temperature. The placental circulation is kept intact while an airway is established.



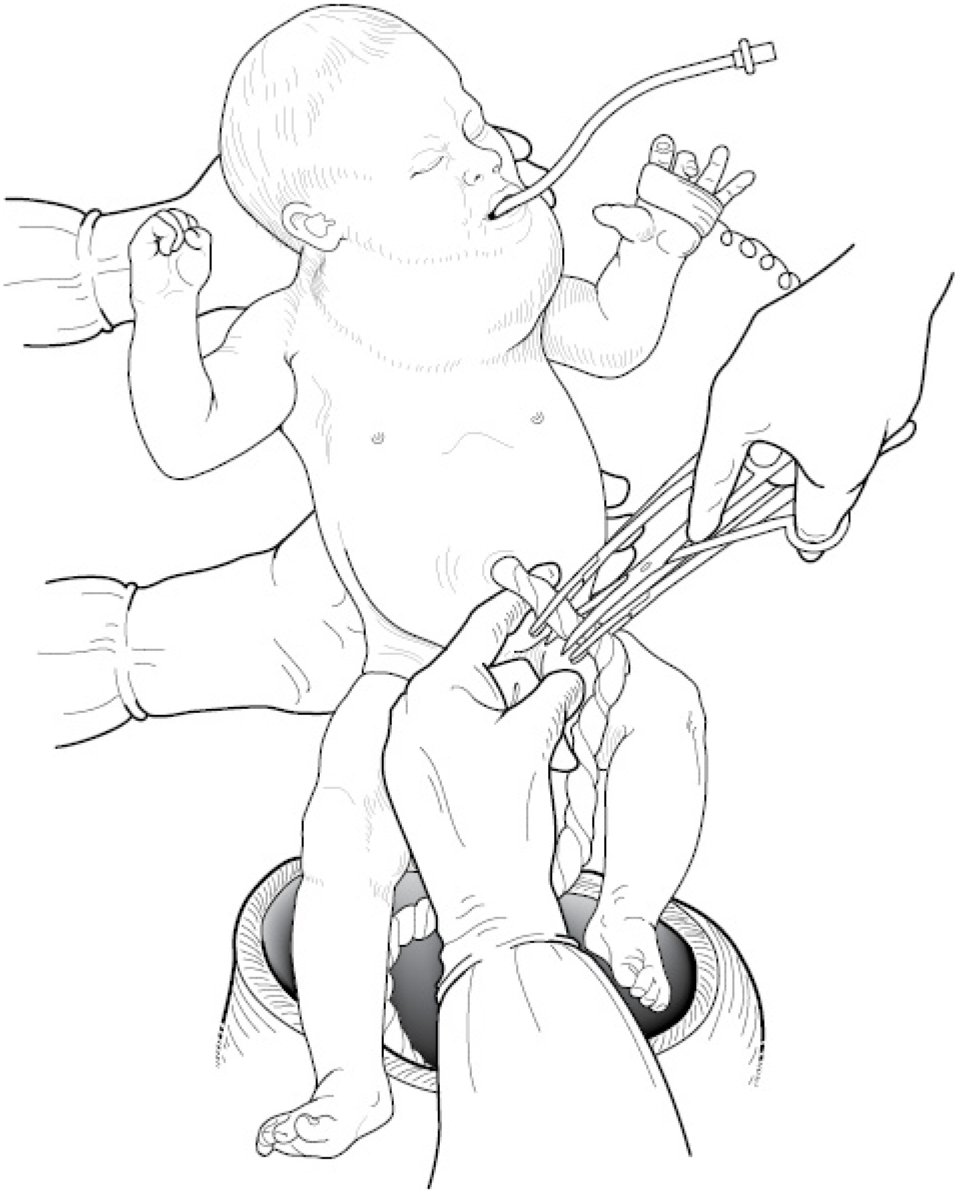
86.7

Irrespective of the procedure, the fetal airway should be controlled as early as possible. Direct laryngoscopy and endotracheal intubation are preferred, but rigid bronchoscopy and/or tracheostomy may be required, keeping in mind that tracheal anatomy may be significantly distorted in cases of giant fetal neck masses, and the carina may be superiorly displaced (**Figure 86.8**). Multiple sizes of laryngoscopes, endotracheal tubes, tube exchangers, as well as rigid and flexible bronchoscopes, should be available.

Intubation may be accomplished either ante- or retrograde, using Seldinger technique through a limited neck dissection. If the airway cannot be accessed by any of these methods, as in the case of a giant neck mass, aspiration or resection should be undertaken. If an airway cannot be established by any means, or in cases where EXIT is performed to avoid hypoxemia prior to ECMO cannulation, ECMO cannulation may be directly performed. Ventilation is initiated prior to cord division once the airway is secure or the operative procedure completed (**Figure 86.9**).



86.8



86.9

Upon completion of the procedure and prior to clamping the cord, umbilical arterial and venous catheters may be placed. Oxytocin should be administered to the mother immediately before delivery, and the child is taken to the resuscitation table once the cord is clamped and divided.

POSTOPERATIVE MATERNAL CARE

Because preterm labor is a relatively common complication after maternal–fetal surgery as well as fetoscopy, in addition to the tocolytic regimen; see "Operation: Exit procedure", uterine activity is monitored by tocodynamometer, and fetal heart rate is followed for any signs of distress. Pain management is continued using the epidural catheter placed preoperatively, which prevents uterine irritability once the inhaled anesthetic has worn off.

Daily fetal echocardiography is performed to evaluate for evidence of indomethacin toxicity, which may manifest as ductal constriction, tricuspid regurgitation, or oligohydramnios. A tocodynanometer is used to monitor uterine activity, and the fetal heart rate is followed for signs of distress. US is also performed daily while the patient is in the hospital to assess amniotic fluid and membrane status, anatomic status, and fetal movement. During the postoperative period, fluid status must be carefully managed due to the risk of non-cardiogenic pulmonary edema. Empiric furosemide diuresis can be added if signs of pulmonary edema develop.

Most maternal–fetal surgery patients are discharged by postoperative day 4, but they should remain on modified bed rest for 2 weeks following discharge. Patients can return to moderate activity after that if no signs of uterine irritability are present. Patients should return for US checks twice per week until delivery. At 36 weeks gestation, an amniocentesis is performed to assess fetal lung maturity, and the fetus is delivered via Cesarean section once the lungs are mature.

COMPLICATIONS

There have been no maternal deaths reported following open maternal–fetal surgery to date, but these procedures are associated with significant short-term morbidity. The most commonly observed complications include preterm premature rupture of membranes, preterm labor, and preterm delivery. Additional complications that have been observed include chorioamnionitis, placental abruption, pulmonary edema, and bleeding requiring transfusion. The overall complication rate is significantly lower for percutaneous procedures. Subsequent pregnancies after open maternal–

fetal surgery are at risk for uterine dehiscence and rupture if labor is allowed to occur, but fertility is not affected.

OUTCOME

Fetal surgery has seen dramatic progress in the last three decades, especially in the ability to diagnose, appropriately select, and treat fetuses with structural malformations that, if left untreated, would result in fetal demise or severely affect quality of life. In some cases, fetal surgery has clearly altered the natural history of the disease and improved outcomes, namely MMC, TTTS, and congenital lung lesions unresponsive to maternal corticosteroid therapy.

The MOMS trial was powered to recruit 200 participants, but was halted after randomization of 183 patients when a planned interim analysis demonstrated clear benefit for prenatal surgery. The fetal surgery group showed significant reduction in rates of shunt placement at 1 year (40% versus 82%) and improvement in neuromotor function by 30 months of age, including the ability to walk without orthotics (42% versus 21%). The degree and presence of hindbrain herniation was also improved, with no hindbrain herniation in 36% of fetal surgery patients and 4% of postnatal surgery patients, and severe hindbrain herniation in 6% of fetal surgery patients and 22% of postnatal surgery patients. The benefits of fetal repair outweighed the complications related to prematurity and the maternal morbidity seen in the study.

The Eurofetus trial was a multicenter randomized controlled trial that compared serial amnioreduction to SLPC for TTTS. The laser therapy group had higher survival of at least one fetus to at least 28 days of age, 76% versus 56% in the amniocentesis group. In addition, the laser group had a higher mean gestational age at delivery, with an average of 33 versus 29 weeks in the amniocentesis group. Most importantly, at 6-month follow-up, the laser group had improved neurologic outcomes, with a decreased risk of periventricular leukomalacia.

Open fetal surgical resections for microcystic CCAM are associated with over 60% survival, with most patients enjoying a good quality of life.

Thoracoamniotic shunt placements for macrocystic CCAM have been reported to decrease CCAM mass volumes by an average of 50% and up to 80%, and are associated with approximately 75% survival.

The benefits of tracheal occlusion in the setting of CDH remain unproven, but a contemporary multicenter randomized controlled trial is currently underway and is expected to lend further insights into the benefits associated with this intervention.

FURTHER READING

Adzick NS. Open fetal surgery for life-threatening anomalies. *Semin Fetal Neonatal Med* 2010; 15: 1–8.

Adzick NS, Thom EA, Spong CY, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med* 2011; 364: 993–1004.

Deprest J, Jani J, Lewi L, et al. Fetoscopic surgery: Encouraged by clinical experience and boosted by instrument innovation. *Semin Fetal Neonatal Med* 2006; 11: 398–412.

Deprest JA, Flake AW, Gratacos E, et al. The making of fetal surgery. *Prenat Diagn* 2010; 30: 653–67.

Golombeck K, Ball RH, Lee H, et al. Maternal morbidity after maternal-fetal surgery. *Am J Obstet Gynecol* 2006; 194(3): 834–9.

Harrison MR, Golbus MS, Filly RA, et al. Fetal surgery for congenital hydronephrosis. *N Engl J Med* 1982; 306: 591–3.

Leichty K. Ex-utero intrapartum therapy. *Semin Fetal Neonatal Med* 2010; 15: 34–9.

Moldenhauer JS. Ex utero intrapartum therapy. *Semin Pediatr Surg* 2013; 22(1): 44–9.

Senat MV, Deprest J, Boulvain M, et al. Endoscopic laser surgery versus serial amnioreduction for severe twin-to-twin transfusion syndrome. *N Engl J Med* 2004; 351: 136–44.

Wilson RD, Lemerand K, Johnson MP, et al. Reproductive outcomes in subsequent pregnancies after a pregnancy complicated by open maternal–fetal surgery (1996–2007). *Am J Obstet Gynecol* 2010; 203: 209.e1–e6.

87

Kidney transplantation

JOHN MAGEE

HISTORY

The first successful kidney transplant was performed between identical twins by Joseph Murray and his colleagues at the Peter Bent Brigham Hospital in 1954. In 1983, the approval of cyclosporine by the Food and Drug Administration (FDA) ushered in the modern era of transplantation. Over the following decades, improvements have continued in all facets of care including pre-transplant management, refinements in surgical technique, improvement in donor selection, perioperative management, and immunosuppression. Currently, there are approximately 750 pediatric kidney transplants performed each year in the United States.

PRINCIPLES AND JUSTIFICATION

Renal transplantation is the treatment of choice for children with end-stage renal disease as it offers the best prospect for health, growth, and development. Absolute contraindications are rare and include ongoing infection and untreated malignancy. Relative contraindications include severe systemic disease that profoundly limits the patient's lifespan or a social situation that makes follow-up with post-transplant care and an immunosuppression regimen absolutely impossible.

Concern regarding the appropriate size for a pediatric recipient relates to the ability to fit the donor kidney within the abdomen, the ability to adequately perfuse a kidney from a significantly larger person, and technical constraints related to any caliber mismatch of donor and recipient's vessels that impacts on the construction of the vascular anastomosis. Accordingly, any target size reflects both recipient size as well as the size of potential donors. Typically, this requires the infant to reach a weight of 6–7 kg, but the principle should be to optimize the situation as far as possible and not let an arbitrary weight target compromise the health of the child.

PREOPERATIVE ASSESSMENT AND PREPARATION

Assessment

All children with chronic kidney disease should be evaluated by a multidisciplinary pediatric transplant team including a transplant surgeon, pediatric nephrologist, social worker, nurse specialist, nutritionist, and psychologist. Efforts to optimize nutrition and dialysis therapy are essential to optimize the child's overall health. When possible, upcoming childhood immunizations should be administered prior to transplantation.

Pediatric candidates are much more likely to have anomalies of the lower urinary tract compared to adults, reflecting the higher incidence of obstructive uropathy or congenital genitourinary abnormalities. An evaluation of the lower urinary system should be completed in such patients, and expertise in these issues or close collaboration with pediatric urology is critical. In general, the patient's native bladder is the preferred reservoir and will usually be adequate. Select patients may require bladder augmentation or other interventions prior to transplantation.

In smaller children with a history of femoral vein catheterization or a history of intra-abdominal process associated with inflammation (e.g. peritonitis, pancreatitis), it is prudent to image the vena cava and the iliac veins to confirm their patency.

It is also important to evaluate and optimize the overall psychosocial state of the child and caregivers. The stress associated with care of a child with a chronic illness is significant, and adherence to the post-transplant medical regimen is vital for success. Adolescents can be particularly challenging in this regard.

Anesthesia

General anesthesia for renal transplantation follows the principles for major abdominal procedures in children. Attention to intraoperative fluid management and maintenance of core body temperature are particularly critical. In children less than 20 kg, the author establishes central venous access, both for fluid administration and monitoring central venous pressure. In these smaller children, the authors also place an arterial line for constant blood pressure monitoring. In larger children, two large-bore peripheral IV lines are sufficient. It is critical to be mindful of the patient's volume status on arrival to the operating room as many patients are polyuric and may be hypovolemic if hydration is not maintained preoperatively.

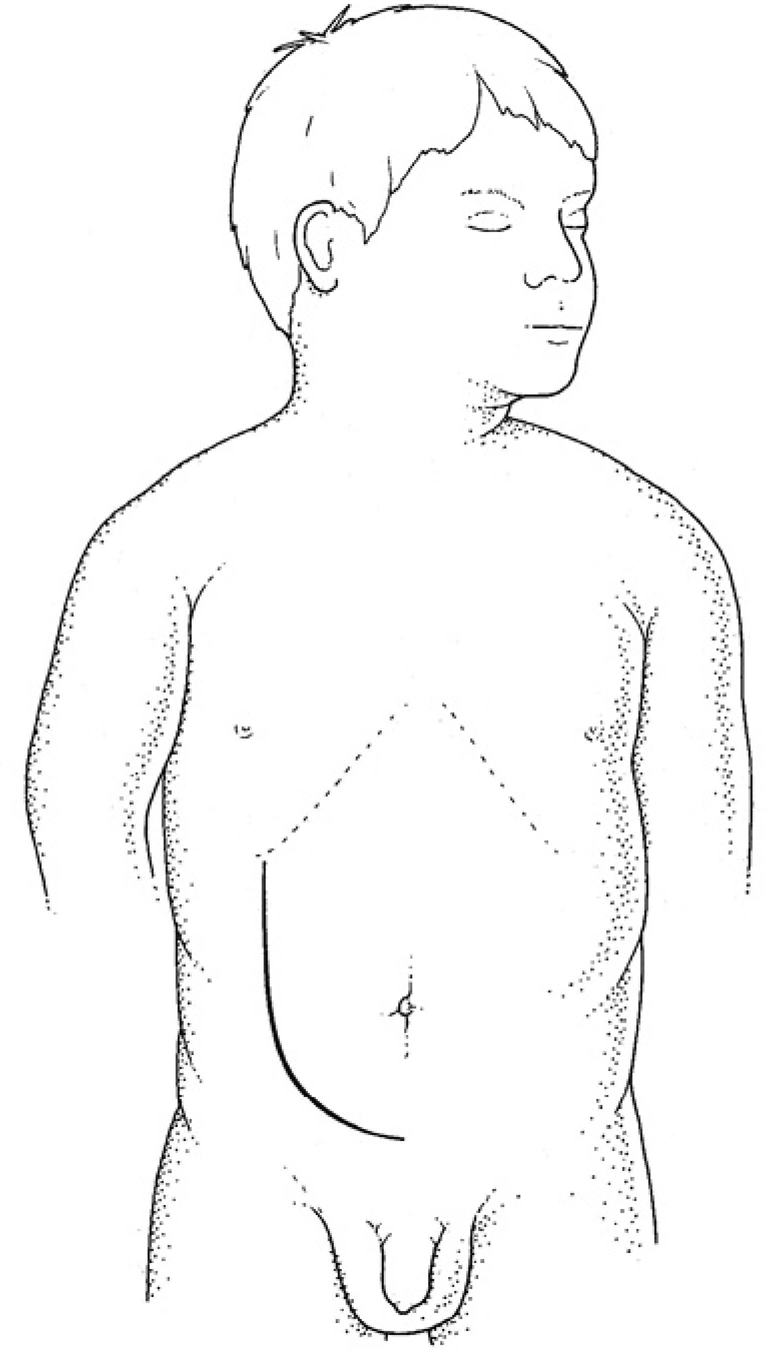
OPERATION

Position of patient

Renal transplantation is performed with the patient in the supine position. A prophylactic antibiotic with coverage against skin flora is administered. A Foley catheter is placed into the bladder or urinary reservoir and connected to a three-way system to allow for instillation of povidone–iodine (Betadine®) solution to distend the bladder at the time of urinary reconstruction.

Incision

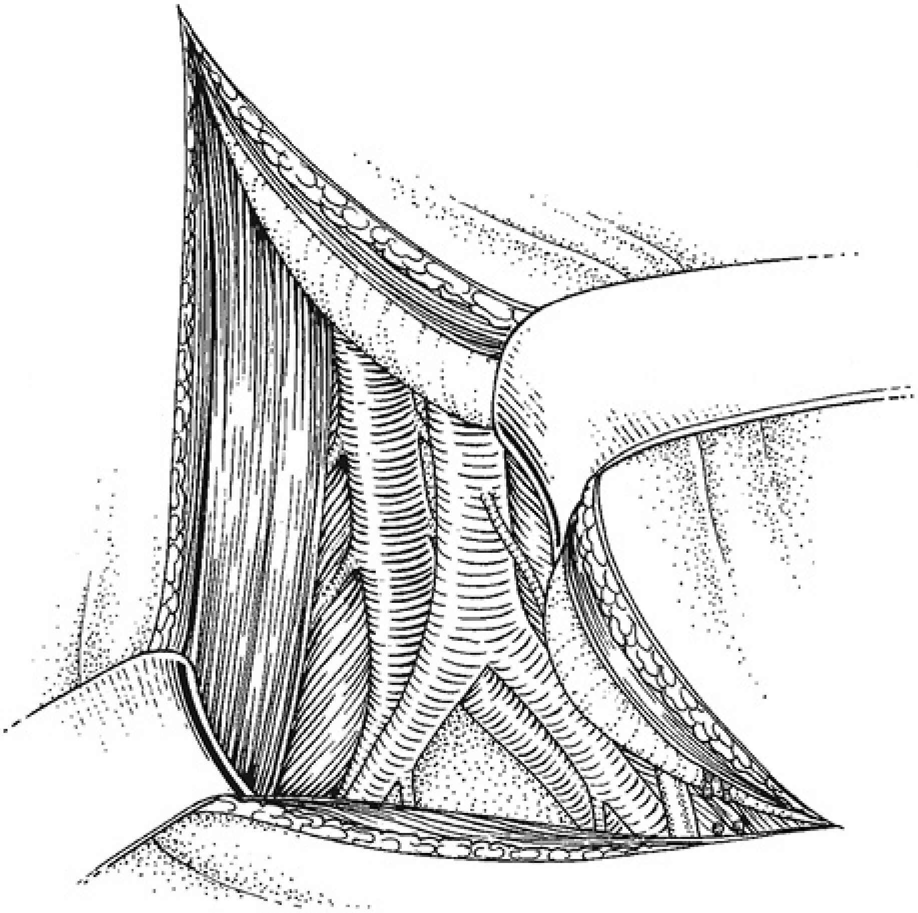
The author utilizes a retroperitoneal approach in all children, including small infants, and prefers to place the kidney on the right side, as this provides easiest access to the vena cava. A curvilinear incision is made, starting at the midline approximately one finger-breadth superior to the pubic symphysis, extending laterally across the rectus abdominis muscle, and then curving superiorly to about the level of the umbilicus. In small children and infants, the incision must be carried further cephalad to permit adequate exposure as the donor kidney is typically from an adult or a larger adolescent (**Figure 87.1**). The significant size mismatch between donor and recipient mandates constant attention to the recipient's hemodynamic profile.



87.1

Exposure

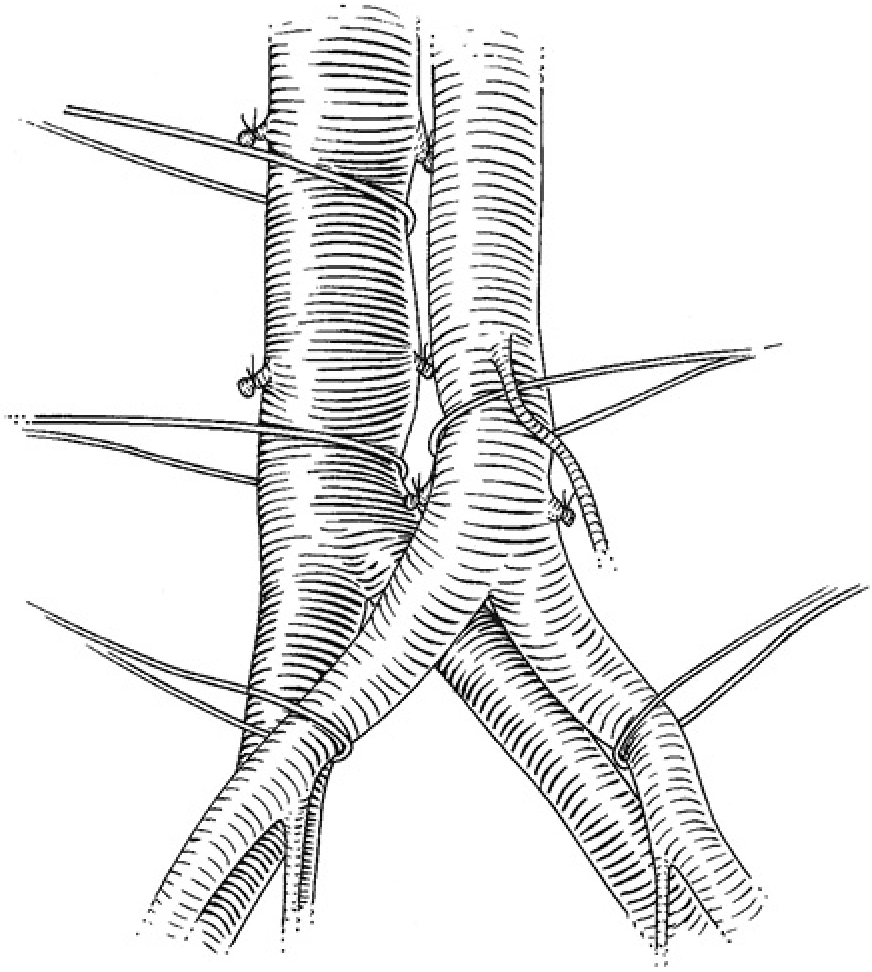
The external oblique, internal oblique, and transversus abdominis muscles are divided, as is the rectus abdominis medially. The preperitoneal space is entered, and the inferior epigastric vessels are typically divided. The peritoneum is swept medially and superiorly to expose the iliac vessels, inferior vena cava, and distal aorta (**Figure 87.2**). Care is taken to avoid inadvertent entry into the peritoneal cavity. Any openings in the peritoneum should be noted and closed at the end of the procedure with absorbable sutures to prevent leakage of peritoneal dialysis postoperatively. The spermatic cord should be preserved by careful mobilization and medial retraction. In girls, the round ligament should be divided. At this point, a self-retaining Bookwalter is placed to provide exposure.



87.2

Vascular dissection

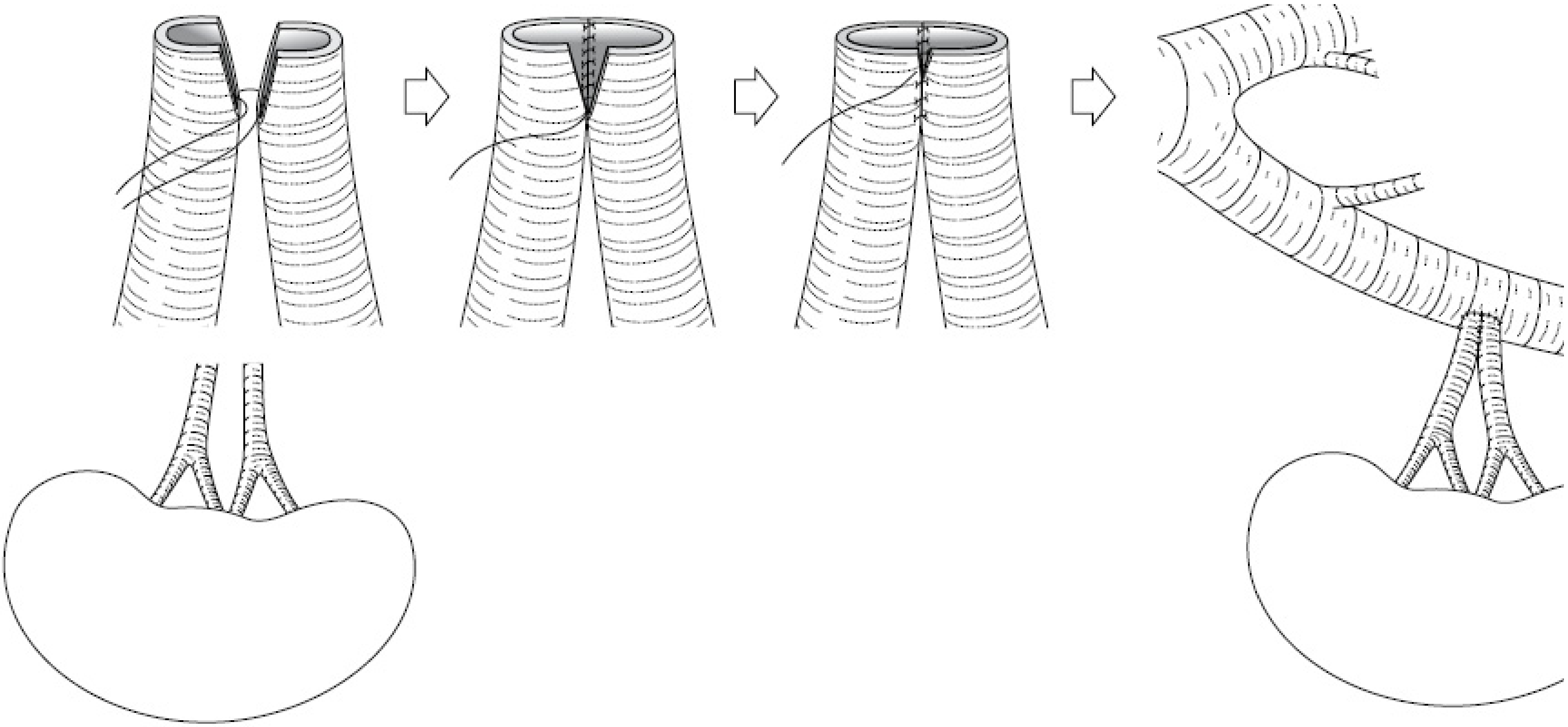
The distal inferior vena cava and aorta are used for vascular reconstruction in children weighing less than approximately 20 kg. Dissection of the distal vena cava is accomplished first. Lumbar veins are divided as needed between fine silk ligatures to free a segment of vena cava approximately 2–3 cm in length. The distal aorta below the inferior mesenteric artery and proximal common iliac arteries are controlled with rubber vessel loops. At least one set of lumbar arteries is generally encountered. These vessels are left intact and controlled with Pott's ties as needed (**Figure 87.3**). Care should be exercised during dissection of the aorta to avoid disruption of the abdominal lymphatic trunk. In larger children, the recipient common iliac vessels may be utilized. In choosing the site for the anastomoses, thoughtful consideration needs to be given for the fit of the kidney in the recipient. Particular attention needs to be focused on the length of the renal vessels and their orientation, considering the ultimate position of the kidney after it is perfused, the retractor is removed, and the fascia are closed.

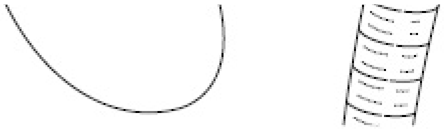


87.3

Preparation of donor kidney

After the recipient dissection is completed, the donor kidney is prepared in a bath of sterile iced saline solution. The renal artery and vein are cleaned of surrounding tissues and side branches are resecured as needed. The renal vein must be kept short, usually not longer than 1–1.5 cm, to prevent kinking. While it is possible to use a Carrel patch of donor aorta if the kidney is from a deceased donor, in practice this usually requires leaving the renal artery too long. Consequently, the renal artery is generally transected to keep it short as well (**Figure 87.4**). If multiple renal arteries are present, they are typically syndactylized before reimplantation, although they can be implanted separately if necessary. When the vessels are syndactylized, it is important to consider if this will allow the vessels to lie in good position, since syndactylization will fix the vessels relatively firmly in two dimensions. This can limit the options of where the anastomosis can be suitably performed or lead to kinking of one or both of the donor arteries if the final position of the kidney is not anticipated. The kidney may be wrapped in a moistened laparotomy pad with a “keyhole” fashioned for the vessels. This facilitates keeping the kidney cool as well as handling of the kidney in the wound.



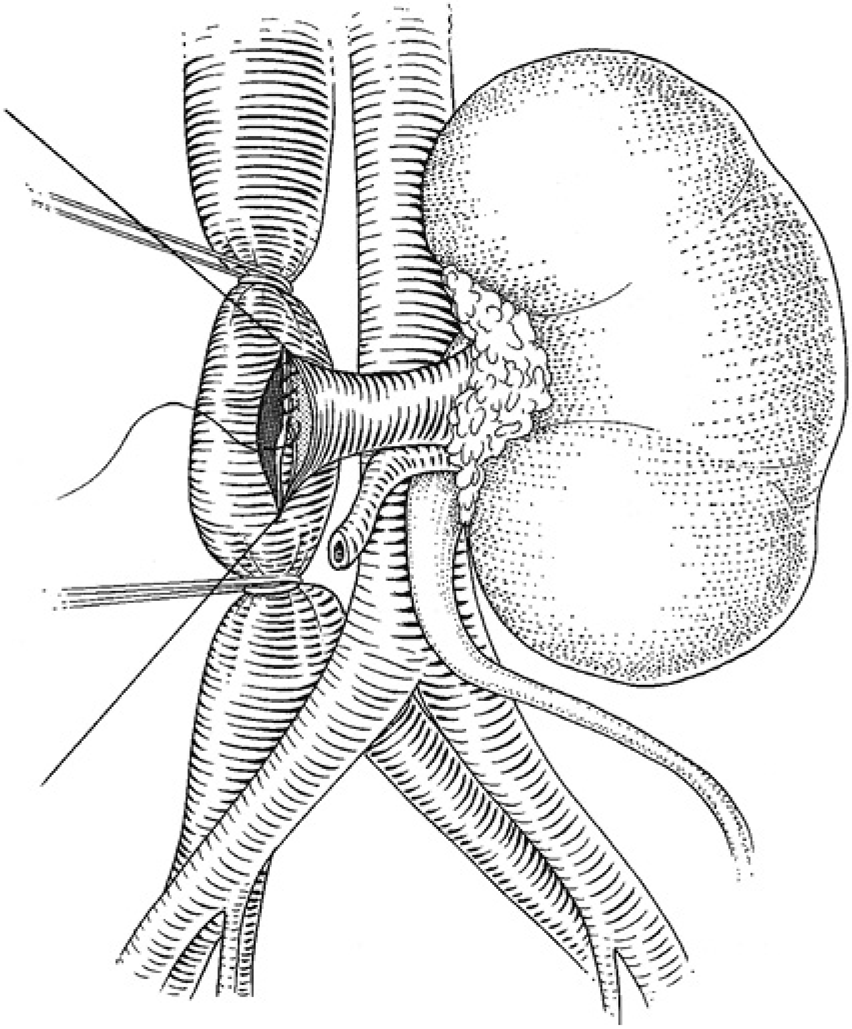


87.4

Vascular anastomoses

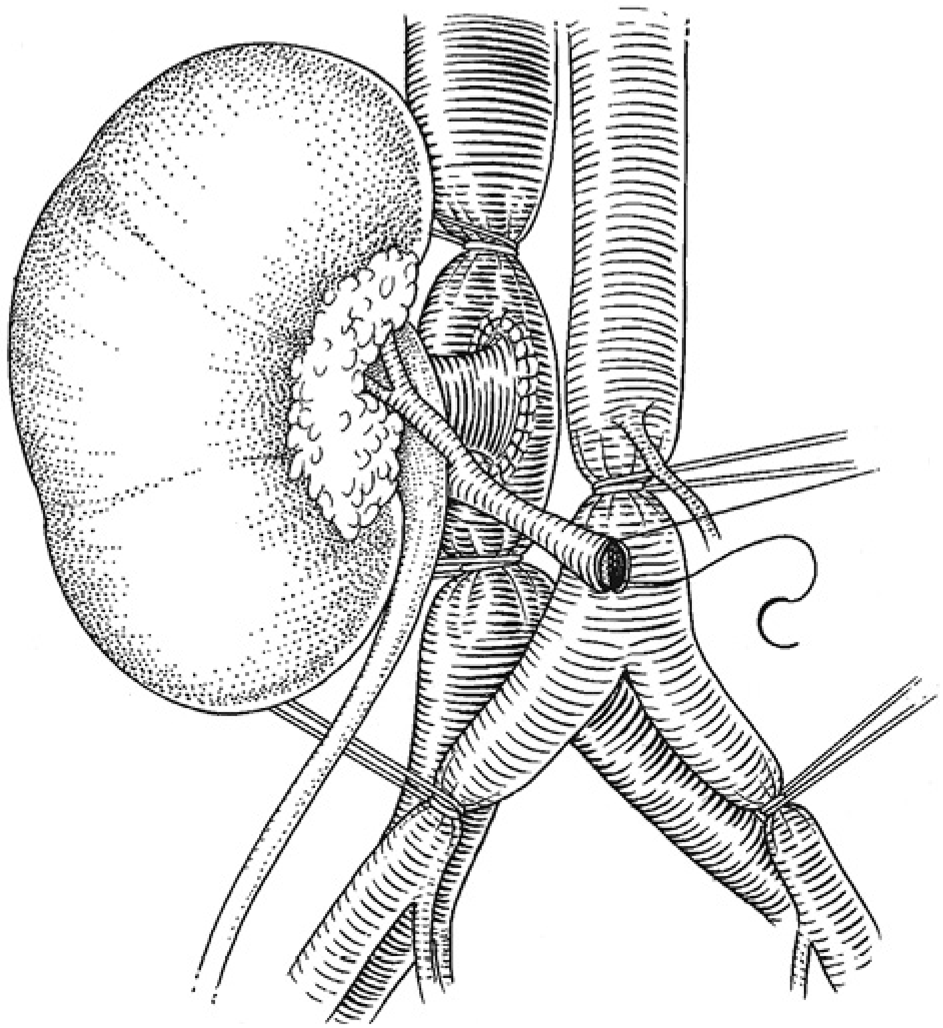
The authors do not routinely heparinize most recipients, though they selectively heparinize children who weigh less than 20 kg receiving deceased donor grafts. In these select cases, the recipient is given 50 units/kg body weight. Full-dose heparinization is unnecessary, except in cases of known hypercoagulability. Vascular clamps should be carefully chosen to avoid obscuring the field. In infants, tension on double-looped rubber vessel slings is all that is required. Alternatively, low-profile gentle vascular spring clips can be used. During construction of the vascular anastomoses, the anesthetic team should volume load the recipient to a central venous pressure of 12–18 cm H_2O and administer mannitol, 0.5 g/kg. These maneuvers will counteract the effects of revascularization and its attendant destabilizing effect due to volume shift. A transplanted adult kidney may sequester 20% of the circulating blood volume of a 7 kg infant.

A longitudinal incision is made in the anterolateral aspect of the vena cava and 5/0 or 6/0 polypropylene sutures are placed at the apices. The assistant positions the kidney medially in the wound. The anteromedial side of the anastomosis is performed first from within the lumen of the vein (**Figure 87.5**), followed by the posterolateral side from the outside. Following completion of the venous anastomosis, the kidney is rotated toward the side of the operating surgeon. In small recipients, the author often places the kidney as it will rest *in situ* from the start, rather than suspend it medially. In this case, the posterolateral side of the anastomosis is performed first from within the lumen.



87.5

The aorta and common iliac arteries are occluded and an aortotomy is made at a site carefully chosen for optimal geometry of the vessels. A 4 mm aortic punch is used to fashion an orifice on the anterolateral aspect of the aorta. This anastomosis is performed with a running 6/0 polypropylene suture, starting at the superior aspect (**Figure 87.6**). The “back wall” of the anastomosis is performed from the inside and the “front wall” from the outside.



87.6

Once the arterial reconstruction is completed, venous outflow is established. The renal artery is then gently occluded with vascular pickups, while restoring distal arterial flow. After several cardiac cycles, inflow to the kidney is established. The kidney should be carefully inspected. Optimally, a uniform pink color and normal turgor are followed promptly by the production of urine. Engorgement of the kidney suggests venous obstruction. This may be due to an imperfect anastomosis, but it is more commonly attributable to compression of the vena cava by one of the retractor blades. A thrill in the renal artery should be developed over time. It is helpful to evaluate flow in the renal parenchyma with a hand-held Doppler probe, paying special attention to evaluating flow in diastole.

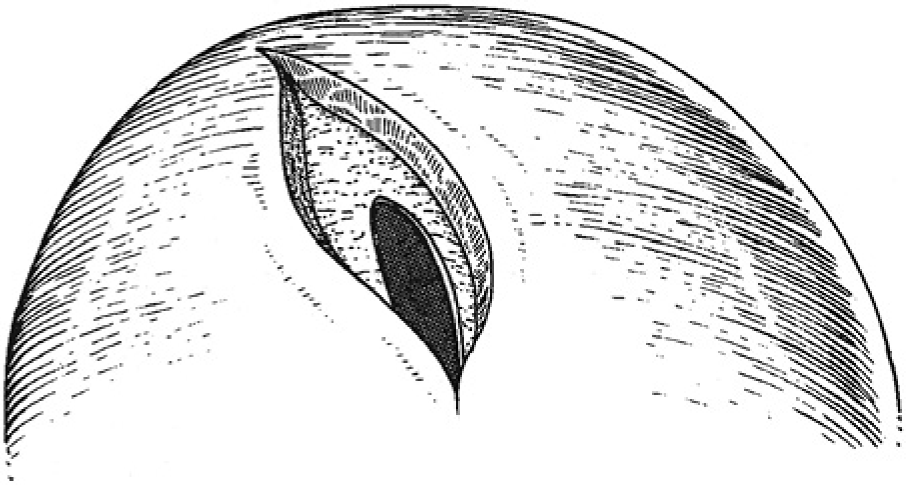
Heparin need not be reversed with protamine sulfate unless troublesome bleeding occurs. Following establishment of hemostasis and assuming satisfactory appearance of the kidney, attention is then turned to the urinary reconstruction.

External ureteroneocystostomy

When the bladder is to be used for urinary reconstruction, the urinary catheter is clamped and the bladder distended by instilling povidone–iodine solution. The author's preferred technique is an external ureteroneocystostomy. The principles of this technique include direct anastomosis of the ureter to bladder mucosa and construction of a submuscular tunnel of sufficient length to prevent reflux into the transplanted kidney.

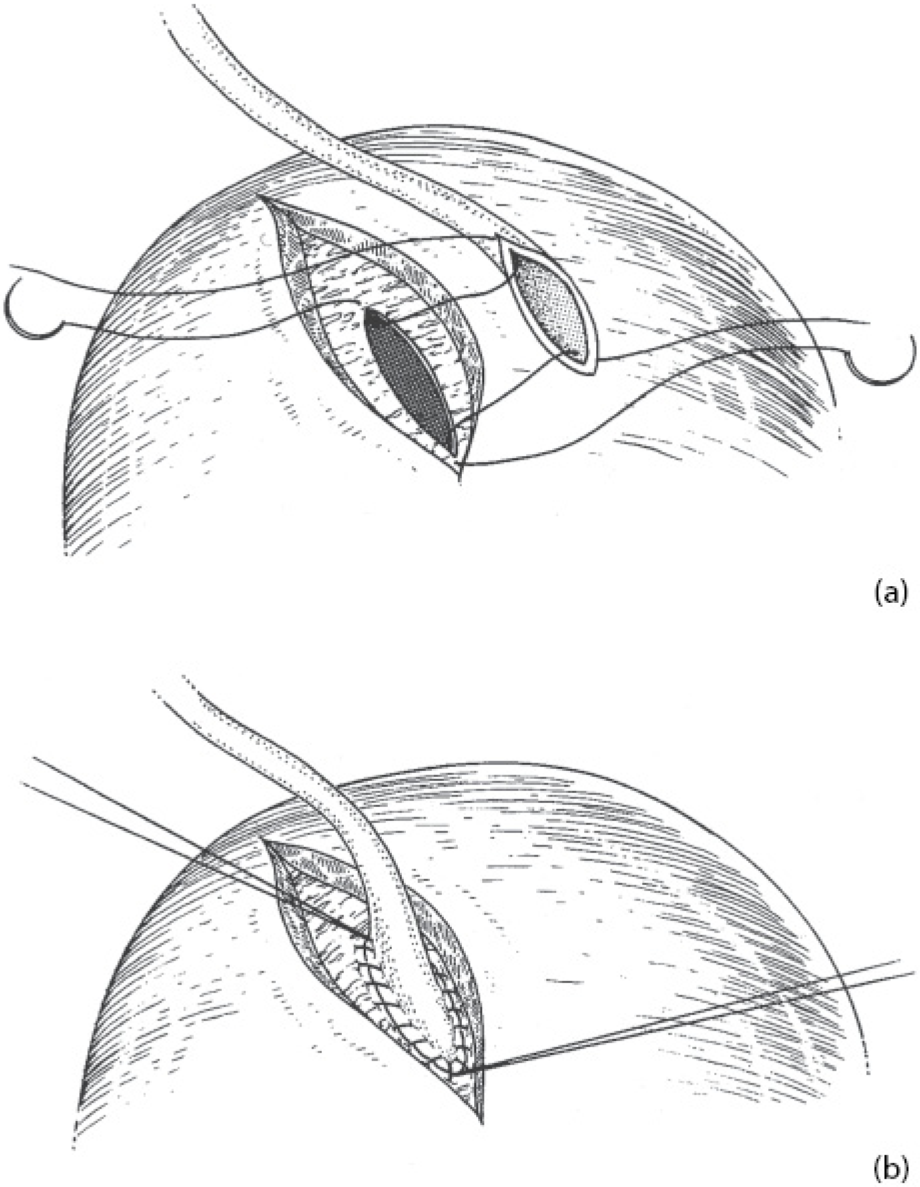
A site is chosen anteriorly near the dome of the distended bladder. The choice of site should take into account the course of the transplanted ureter. An appropriate balance must be struck between the need for an adequate ureteric length to reach the anastomotic site when the bladder is empty and the requirement of avoiding excessive redundancy with the attendant risk of ureteric obstruction and distal ureteral ischemia. Given the blood supply to the transplanted ureter is dependent on small branches from the lower-pole renal artery, the ureter should be kept as short as possible. In males, the ureter should be brought beneath the spermatic cord to avoid obstruction caused by draping the ureter over the cord.

The muscular coat of the bladder is divided, exposing the bladder mucosa for a distance of about 4 cm. An opening is made in the mucosa at the medial aspect of the dissection (**Figure 87.7**). The presence of the bladder catheter should be confirmed visually at this time; a thickened peritoneum adjacent to the bladder can fool even the most experienced surgeon.



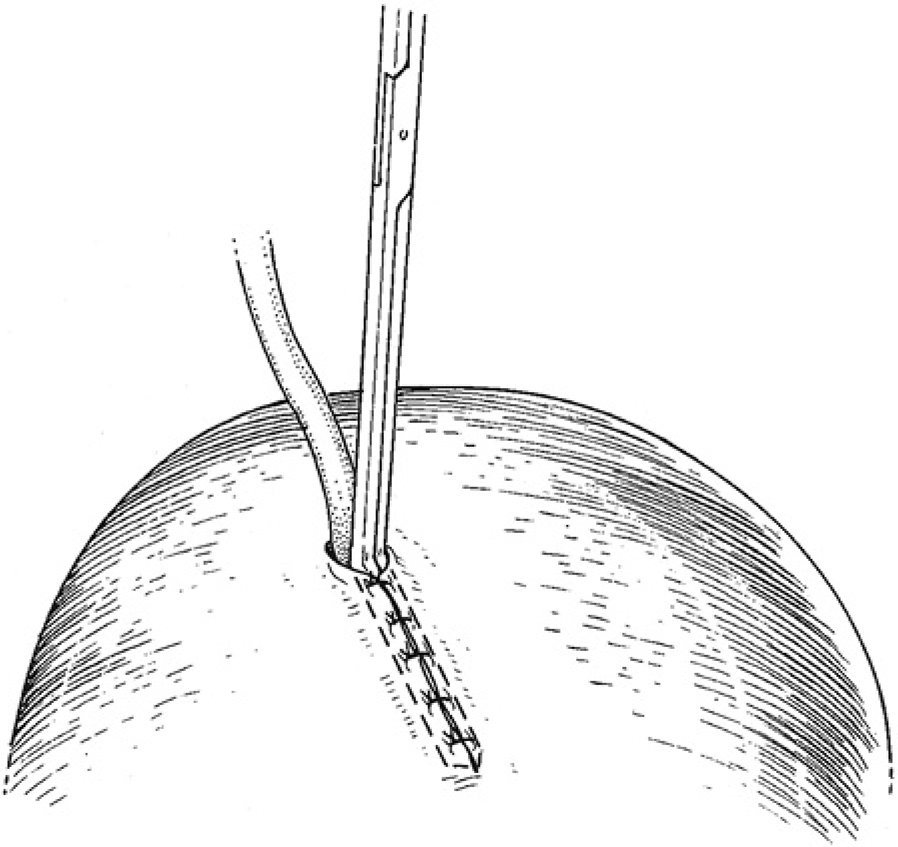
87.7

The ureter is cut to length and spatulated. The anastomosis is performed with running 5/0 monofilament absorbable sutures. Traction sutures are placed at the “heel” and “toe” of the anastomosis (**Figure 87.8a**). Construction of the anastomosis should proceed from “heel” to “toe” on the inferior wall and then again “heel” to “toe” on the superior wall (**Figure 87.8b**). This permits precise placement of sutures at the “heel” where the opportunity for compromising the lumen of the ureter is greatest. Precise attention must be focused on the tension applied to the suture during follow-through and while tying the knot in order to avoid cinching the suture line, which would narrow the anastomosis. Careful placement of sutures and avoidance of excessive handling of the ureter are critically important to avoid stenosis or obstruction. The authors routinely place a double-J stent after the anastomosis is partially completed, with the upper end positioned in the renal pelvis and the lower end passed into the bladder.



87.8a,b

After completion of the anastomosis, the ureter is laid in the submuscular space and the muscle wall is approximated over it using interrupted 4/0 monofilament absorbable sutures for a distance extending 2–3 cm beyond the anastomosis. The overall length of the tunnel compared to the anastomosis should be 4 : 1 to 5 : 1. When completed, the tunnel should still admit the end of a right-angled clamp, thus ensuring that the ureter will not be obstructed within the tunnel (**Figure 87.9**).



87.9

Wound closure

The transplanted kidney should lie comfortably within the lower abdomen. No fixation of the kidney allograft is needed. On removal of the self-retaining retractor, the peritoneal contents will hold the kidney against the posterior and lateral side walls. No drains are used. The wound is irrigated and a one-layer fascial closure is completed with running absorbable suture. The skin is closed with a running, absorbable, subcuticular suture.

POSTOPERATIVE CARE

General care and infection prophylaxis

Infants and small children are managed in a pediatric intensive care unit. Although most recipients are extubated in the operating room, a small infant with an adult-sized kidney may require mechanical ventilation for 1–2 days. Urine output is replaced with IV fluid milliliter for milliliter for the first 12 hours. Replacement fluids are then gradually tapered. Maintenance of adequate volume status is critical. Any concern regarding graft function should be promptly evaluated via ultrasound with Doppler. The urinary catheter is left in place for 2–3 days. If necessary, satisfactory bladder emptying may be assured with post-micturitional catheterization.

In our center we do not administer additional antibiotic prophylaxis postoperatively. Prophylaxis is given for 90 days against opportunistic infection including trimethoprim and sulfamethoxazole for *Pneumocystis carinii*, and valganciclovir for *cytomegalovirus* infection. Nystatin is administered for 30 days as prophylaxis for candidiasis. The hospital stay is 6–12 days in most cases.

Immunosuppression

Over the last decade, the introduction of several new agents has permitted several protocol permutations. Immunosuppression protocols tend to be center-specific, reflecting local and national experience, and a desire to appropriately balance the risk of rejection with the risks of immunosuppression. Induction immunosuppression is less common in countries where the population is more homogeneous and the risk of rejection appears lower. Accordingly, dual-agent maintenance therapy is also more common in such settings. Immunosuppressive therapy starts intraoperatively. The author uses induction therapy with polyclonal anti-thymocyte antibody (Thymoglobulin®) for recipients at higher immunologic risk and anti-interleukin-2 receptor monoclonal antibody (basiliximab) for all others. Corticosteroids are also administered intraoperatively. Postoperatively, the calcineurin inhibitor tacrolimus is introduced when renal function is demonstrated. The anti-metabolite, mycophenolate mofetil, completes the standard triple-drug maintenance regimen. Based on recent experience with several multicenter trials, we offer a steroid avoidance protocol where corticosteroids are stopped after 5 days. For patients who are continued on corticosteroids, the dose is rapidly tapered and most patients are maintained on alternate day steroids.

OUTCOME

The overall results of renal transplantation in children have improved steadily. The most important advances have included improvements in pre-transplant dialysis and nutritional management, meticulous surgical technique, and management of immunosuppression. Transplant and developmental outcomes have been excellent even among recipients less than 1 year of age. Graft survival is dependent on donor type and recipient age. For pediatric recipients of living donor kidneys, 1- and 5-year graft survival ranges are 96–99% and 84–92%, respectively. For deceased donor kidneys, 1- and 5-year graft survival is 94–98% and 77–86%, respectively. Adolescent recipients account for the lower range of graft outcomes at 5 years.

FURTHER READING

Dharnidharka V, Fiorina P, Harmon W. Kidney transplantation in children. *N Engl J Med* 2014; 371: 549–58.

Hebert SA, Swinford RD, Hall DR, et al. Special considerations in pediatric kidney transplantation. *Adv Chronic Kidney Dis* 2017; 24: 398–404.

Riley P, Marks SD, Desai DY, et al. Challenges facing renal transplantation in pediatric patients with lower urinary tract dysfunction. *Transplantation* 2010; 89: 1299–307.

Salvatierra JO, Millan M, Concepcion W. Pediatric renal transplantation with considerations for successful outcomes. *Semin Pediatr Surg* 2006; 15: 208–17.

Van Arendonk KJ, Boyarsky BJ, Orandi BJ, et al. National trends over 25 years in pediatric kidney transplant outcomes. *Pediatrics* 2014; 133: 584–601.

88

Liver transplantation

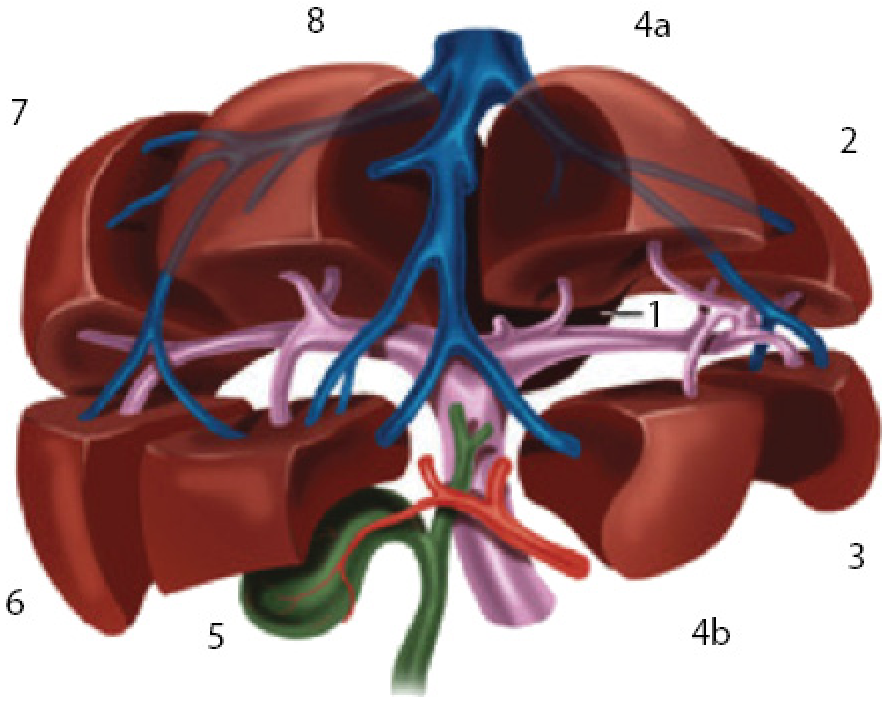
CAROLINE LEMOINE AND RICCARDO A. SUPERINA

HISTORY

A series of three human liver transplants was performed in 1963 by Thomas Starzl and his team in Denver, CO, USA. The first attempt was for a child with biliary atresia, but who did not survive the operation. The others were for adults with liver tumors and were technically more successful, showing the potential of this approach, though attempts at control of rejection with the immunosuppression available were more problematic.

In the pediatric field, subsequent refinement of technique has allowed for the reduction in size of adult donor livers to accommodate the smaller-sized organ needed in infants and children. This has allowed more timely transplantation of a child waiting for an organ, and has decreased waiting-list mortality. Improved immunosuppression with calcineurin inhibitors such as ciclosporin and tacrolimus, together with others such as mycophenolic acid and rapamycin, has improved their long-term survival. More effective antiviral and antibacterial therapy, as well as improved detection techniques for post-transplant lymphoproliferative disease, has also contributed to the improved survival. Lastly, more refined techniques of organ procurement, preservation, and implantation, in addition to better peri- and postoperative care have all contributed to improve outcomes over the past four decades.

The techniques of hepatic size reduction permitting segmental transplantation of the liver, including split, reduced size, and living-donor liver transplantation (LT), require familiarity and understanding of the segmental liver anatomy described by Claude Couinaud (**Figure 88.1**). Reduced liver grafts are prepared by cutting down a deceased donor adult liver to produce a left lateral segment graft (segments 2 and 3), left lobe graft (segments 2, 3, and 4), or more rarely, the right lobe (segments 5–8) and were introduced in 1984 by Henri Bismuth. Split-liver transplantation, in which a deceased donor adult liver is divided into two functional grafts, became available in 1988 after its description by Rudolf Pichlmayer. Pediatric living-donor LT, in which a healthy adult donates part of his or her liver, followed in 1989 after its first successful application in Australia. Living-related donation enabled the development of pediatric liver transplant programs in countries without established deceased donor organ systems, such as Japan and Korea, and has since been extended to both adults and adolescents through the use of right lobe donation.



88.1 Couinaud's liver segments.

Still the greatest barrier to survival for children who need a transplant is donor organ availability. Since most children awaiting transplantation are infants or small children and the peak age in donors is in the adult ranges of age, attempts at finding age- and size-matched organs for children has been associated with a high mortality for those waiting for organs. Size reduction techniques have greatly reduced although not eliminated the waiting-list attrition in lives, particularly in patients under the age of 1 year.

PRE-TRANSPLANT CONSIDERATIONS AND MANAGEMENT

Indications

The main indications for pediatric LT are listed in **Table 88.1**.

Table 88.1 **Indications for liver transplantation**

Acute liver failure	
Viral (e.g. hepatitis A, B, non-A-E) Drugs, toxins, cryptogenic Autoimmune	
Chronic liver disease	
Cholestatic	Biliary atresia Progressive familial intrahepatic cholestasis (PFIC) Alagille syndrome Sclerosing cholangitis
Metabolic	α-1-antitrypsin deficiency Wilson's disease Cystic fibrosis Tyrosinemia Urea cycle defects
Liver tumors	e.g. hepatoblastoma, hepatocellular carcinoma
Autoimmune	

Chronic liver disease with cirrhosis is the most common indication for LT in children, and biliary atresia is the most common form of chronic liver disease leading to transplantation, accounting for approximately one-third of liver transplants in most series. Most children with biliary atresia have already undergone a Kasai portoenterostomy in the first 2–10 weeks of life, with failure characterized by progressive jaundice, increasing portal hypertension, ascites, and malnutrition. Even those with initial clearance of jaundice may still require LT later. Chronic liver disease may also lead to progressive encephalopathy, metabolic bone disease, intractable pruritus, pulmonary hypertension, and hepatopulmonary syndrome. There are no uniformly accurate predictors of success following the Kasai procedure, but some infants will derive no benefit in terms of relief of jaundice and will require LT within 1–2 years of life. Those who are diagnosed late with biliary atresia may benefit from a primary transplant.

Alagille syndrome and progressive familial intrahepatic cholestasis (PFIC) are both inherited cholestatic disorders that may require LT during childhood. Full cardiac evaluation is required before proceeding with LT in those with Alagille syndrome because of associated anomalies. PFIC, caused by mutations in the genes involved in bile acid transport, can be divided into three subtypes depending on clinical features and genetic characteristics. Occasionally, surgical bile diversion procedures are effective at controlling pruritus in those without evidence of end-stage liver disease. However, these appear to be less effective in those with PFIC-2, and this subgroup may benefit from LT. PFIC-1 (Byler's disease) patients, on the other hand, may suffer from intractable diarrhea post-LT.

Also common indications for LT are inborn errors of metabolism resulting from liver enzyme deficiencies or mutations, such as alpha-1-antitrypsin deficiency or tyrosinemia. The latter is no longer considered a primary reason for LT since early detection can lead to successful treatment preventing the most serious manifestation. Wilson's disease is a genetic disorder of copper metabolism with excessive copper deposition in the liver, basal ganglia, kidney, and other organs. Most cases can be successfully treated medically, but some patients present acutely with liver failure and need an urgent LT. Urea-cycle defects and maple syrup urine disease are metabolic disorders that occur in the presence of otherwise intact hepatic synthetic function. These conditions are associated with the development of potentially irreversible neurological sequelae and require strict adherence to dietary modifications and enzyme replacements. LT restores the deficient enzyme and has been shown to improve quality of life. However, transplant does not change neurological conditions once they have occurred. It is therefore recommended to proceed with LT early in infancy.

Acute liver failure (ALF) is not common in children, but the situation may warrant extremely urgent action, and it tests the preparedness of a transplant program to mobilize numerous resources to get a child transplanted. Viral hepatitis is the most common cause for ALF, but the specific virus responsible is rarely identified. Drug-related toxicity and some metabolic liver diseases, such as Wilson's disease, can also cause acute decompensation. In up to 50% of pediatric patients with ALF, no identifiable diagnosis will be made (cryptogenic). ALF, by definition, is the onset of encephalopathy within 8 weeks of the appearance of jaundice, and the progression of encephalopathy can be a slow or rapid response to brain death within 24 hours. ALF is potentially reversible, and the mainstay of treatment is supportive care. Nevertheless, the development of hyperammonemia and hepatic encephalopathy should be treated aggressively, because of the risk of developing cerebral edema and irreversible brain damage. Most programs proceed with listing of the patient for LT with the onset of encephalopathy. Urgent living-donor transplantation has been used in this setting with great success in experienced programs since the wait for a deceased donor organ can be prolonged in a situation where the patient's status may be deteriorating by the hour.

Apart from the onset of encephalopathy, poor prognostic signs include prolongation of the international normalized ratio (INR) above 2.5, non-acetaminophen (paracetamol) related liver injury, age <10 years, and worsening renal function.

Liver tumors, such as hepatoblastoma, that are not safely resectable after neoadjuvant chemotherapy may also be transplanted with good results. The presence of pulmonary metastases at diagnosis does not constitute in itself an absolute contraindication to LT. If lung metastatic disease effectively clears after neoadjuvant chemotherapy, or if it persists but is amenable to complete surgical resection, the patient may be eligible for LT.

Hepatocellular carcinoma occurs less frequently than hepatoblastoma in children, and occurs most often in non-cirrhotic livers, in contrast to adults. It is a more aggressive tumor than hepatoblastoma, is usually found to be chemoresistant, and is associated with a worse prognosis. LT appears to offer a higher survival rate (~70%) than primary resection (30–50%) in children.

Contraindications to LT include persistent extrahepatic malignant disease, severe extrahepatic disease that cannot be reversed by LT (such as severe pulmonary hypertension), uncontrolled systemic sepsis, and severe irreversible neurological injury. Metabolic disorders that affect other organ systems and could lead to death even if the hepatic function was improved are also contraindications.

Timing

Transplantation must be considered in any child with chronic liver disease who is showing signs of decompensated cirrhosis, including growth failure and malnutrition. Portal hypertension alone in a well-compensated cirrhotic may be treated by non-transplant means, but the presence of ascites, non-correctable coagulopathy, hypoalbuminemia, and metabolic bone disease should trigger a transplant evaluation, and quite likely activation on a transplant waiting list.

The pediatric and adult end-stage liver disease scores (PELD and MELD, respectively) were designed to quantify the need for transplantation in an objective manner, and scores >15 are considered as representing a relatively high risk of dying within a year without a liver transplant. Exceptions to this rule are numerous and lower scores should not exclude a child from being evaluated.

It is important to initiate the evaluation and listing for transplantation in a timely fashion that will allow for a donor organ to be found before the recipient's condition deteriorates to the point that success is unlikely. Factors pointing to a poor outcome after transplantation include incompletely treated sepsis, pulmonary disease with high oxygen requirements, requirements for inotropes to maintain an adequate blood pressure for age, and declining renal function.

In children with acute or subacute liver failure, the poor prognostic signs as outlined above should lead to a rapid evaluation of the child for transplantation and once the decision has been made to transplant a child, speed is of the essence since neurological or hemodynamic deterioration may be rapid.

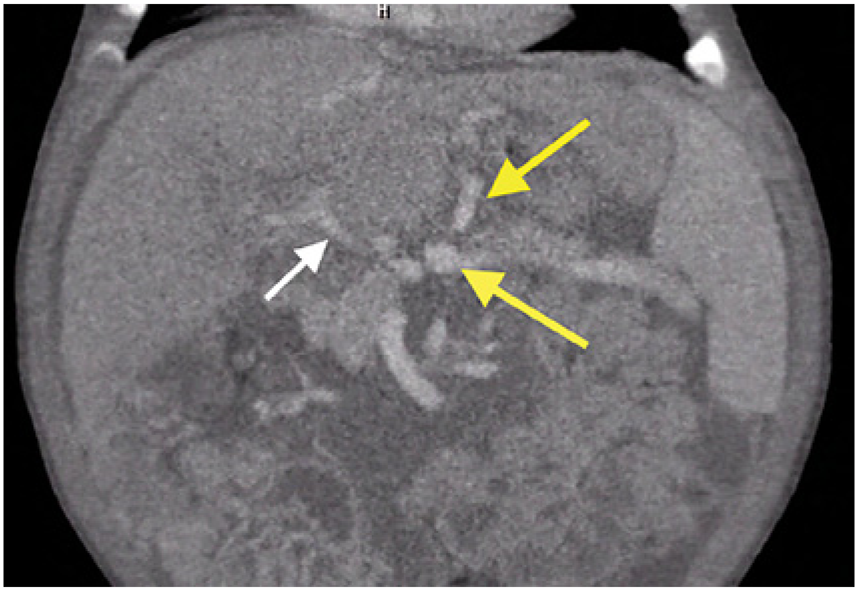
PREOPERATIVE ASSESSMENT AND PREPARATION

The pre-transplant evaluation includes assessment by a multidisciplinary team which may include surgery, anesthesia, hepatology, social work, pharmacy, dentistry, and other medical specialties as needed e.g. cardiology. Specific elements of assessment include:

- Liver anatomy, particularly the status of the portal vein. Abnormality, absence, or stenosis may suggest the need for a vein graft interposition, or using the superior mesenteric vein as the inflow. Arterial and caval anatomy may be less critical but the former is usually very variable and may be specifically abnormal in conditions such as Alagille syndrome.
- Exclusion of comorbidities such as pulmonary hypertension or renal disease.
- Correction of metabolic, nutritional, and immunization deficiencies.
- Dental examination.
- Psychosocial impact of LT on the family with a social worker evaluation.
- Education of the family about the procedure, long-term implications, and sequelae. This should include a discussion about potential living-donor transplantation and making this option available if it is desired.

A broad range of investigations may be indicated but the preoperative workup should be customized based on the underlying cause of liver failure. Malnutrition is common because of malabsorption, anorexia, and catabolism. Nutritional status is a major determinant of outcome. Aggressive nutritional support, including nasogastric or even parenteral nutrition, may be needed. Bleeding esophageal varices are treated, preferably by variceal band ligation. Viral screening should include cytomegalovirus (CMV) and Epstein–Barr virus (EBV) serology, both of which are important potential post-transplant pathogens. Patients should be immunized against *Pneumococcus* and hepatitis A and B in addition to routine vaccines. Live vaccines, such as varicella, are best given at least 30 days before transplant, as they are generally contraindicated in the immunosuppressed patient.

It is essential to define the vascular anatomy of the liver prior to a transplant. Patients with biliary atresia may have a variety of both congenital and acquired vascular problems. A Doppler ultrasound (US) examination looking for portal vein flow and size is essential. A computed tomography (CT) or magnetic resonance angiogram (MRA) may be indicated if other anomalies are suspected. Preoperative demonstration of patent hepatic vasculature is essential to assess the technical feasibility of the operation. US is often sufficient to determining the patency and size of the portal vein and delineate the recipient vascular anatomy. Failure to visualize satisfactory hepatopedal portal blood flow, however, is an indication for CT angiography. In the example shown in **Figure 88.2**, there is a very small portal vein (white arrow) with a large splenic and coronary vein to the patient's left (yellow arrows). In this patient, the surgeon should be prepared to reconstruct the portal vein with a vein graft if necessary.



88.2 Small PV, large SV, and coronary.

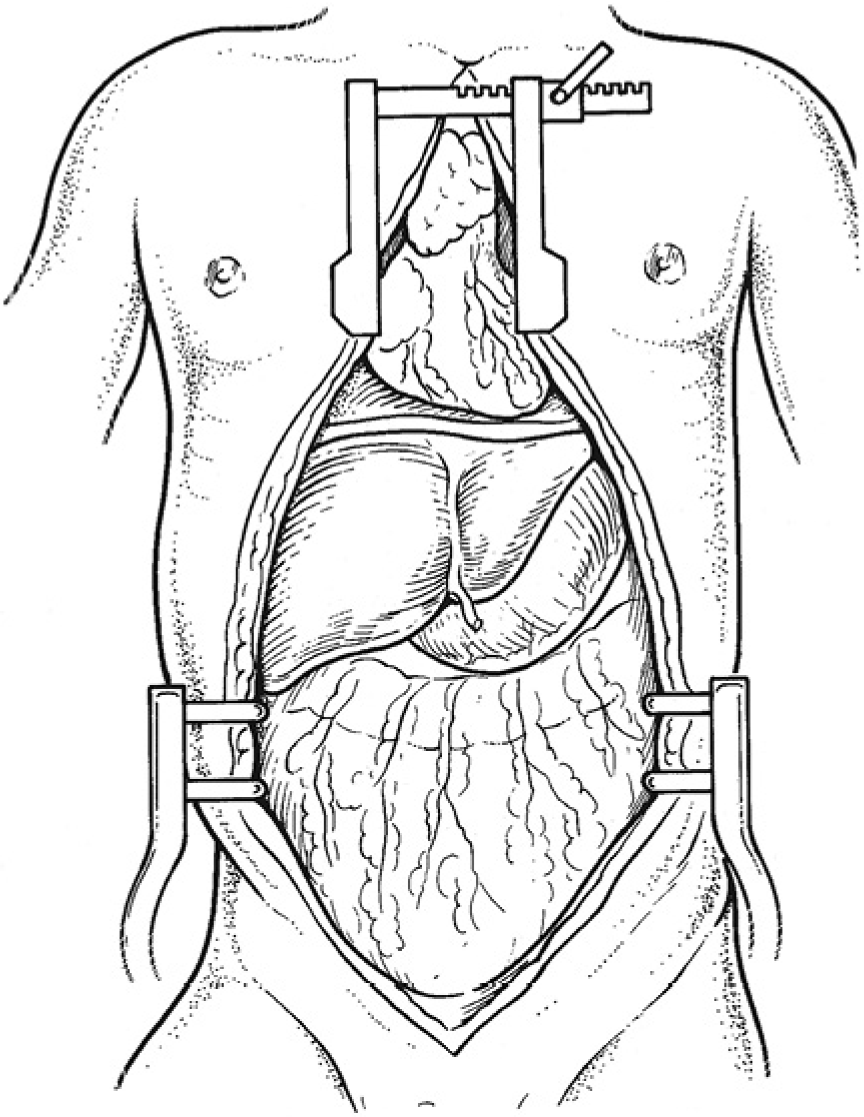
DONOR OPERATION

Although livers from donors following cessation of cardiac function have been used in children, this practice is generally avoided due to the high incidence of ischemic cholangioapthy in the recipient. The liver is typically procured from a heart-beating donor in whom brain death has been diagnosed by stringent criteria. Most donors for pediatric LT are aged between 1 and 50 years and have died from severe head injury, spontaneous intracranial bleed, or treated bacterial meningitis. More cautiously, deaths from intracranial tumors can also be used.

The donor and recipient should be ABO blood group compatible, but tissue typing is not required. ABO incompatible donors are justified in urgent situations, particularly for children below the age of 1 year in whom preformed anti-ABO antibodies are not present.

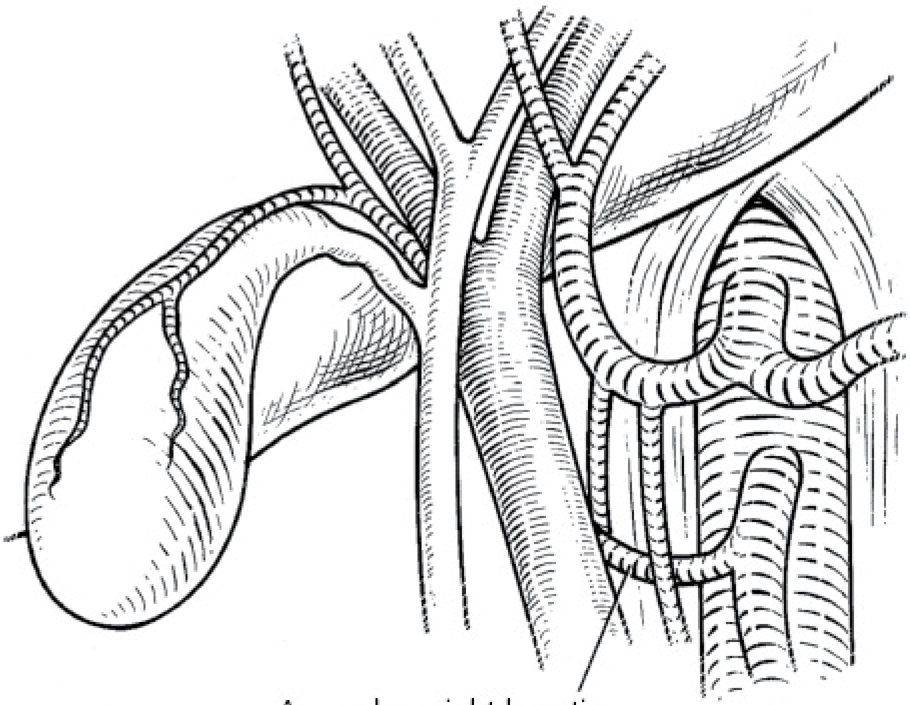
The donor should have near-normal biochemical liver function, no history of extracranial malignancy, and no evidence of chronic liver disease or past infection with hepatitis B or C, human immunodeficiency virus (HIV), or prions. Donor CMV and EBV serology must be recorded. Most donor livers are removed as part of a multiorgan procurement procedure in which the liver, kidneys, pancreas, heart, and/or lungs are removed.

A midline incision is made from the suprasternal notch to the pubis. The sternum is opened in the midline using a sternal saw and retractor (**Figure 88.3**). We routinely preserve the falciform ligament and the associated fat pad attached to the graft as an option to pad a potentially redundant arterial reconstruction and prevent kinking. If not needed, this fat pad is tied and resected following reperfusion. The abdominal viscera are carefully inspected to exclude injury or disease. The liver is mobilized by incising its ligamentous attachments. The gastrohepatic ligament is examined for the presence of an accessory/replaced left hepatic artery arising from the left gastric artery, which, if present, must be preserved in continuity with the celiac axis.



88.3

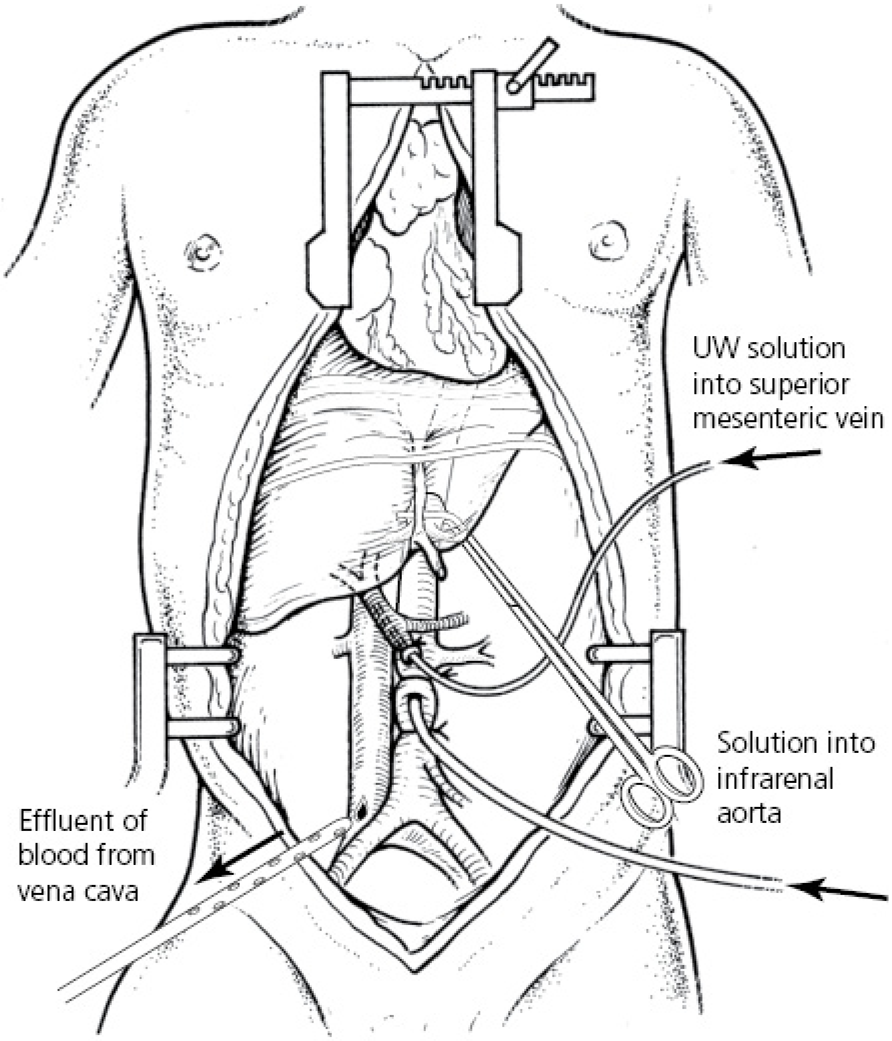
The porta hepatis is dissected next, checking for an anomalous right hepatic artery arising from the superior mesenteric artery (**Figure 88.4**). This usually lies behind the portal vein and bile duct, and must be preserved in continuity with the superior mesenteric artery.



88.4

If there is a conventional hepatic arterial supply, the gastroduodenal and right gastric arteries are divided between ligatures followed by ligation of the left gastric and splenic arteries. The common bile duct (CBD) is divided close to the upper border of the duodenum, the gall bladder is opened and carefully flushed with cold saline. The portal vein is carefully dissected. More rapid techniques do minimal dissection at body temperature to reduce the risks of bleeding or donor hemodynamic instability. Division of arterial structures such as the splenic, left gastric, and gastroduodenal arteries takes place in the cold without tying them off, and these must be dealt with on the back table in the cold when the liver is prepared for implantation or in the recipient before reperfusion.

The distal aorta just above the bifurcation is encircled with two strong tapes and the aorta just above or below the diaphragm is next dissected and encircled with a tape. Once all procurement teams have completed their respective organ dissection, the donor is heparinized. A large-bore cannula is then inserted into the distal aorta and used to flush the viscera with up to 5 liters of any number of available preservation solutions (e.g. University of Wisconsin (UW) solution) (**Figure 88.5**). The common factor in all preservation solutions is that they are cooled to 4 °C prior to use and that they mimic intracellular electrolytes with a high potassium concentration. We do not routinely use a portal vein flush *in situ*. The subdiaphragmatic aorta is clamped, the infrarenal vena cava (or intracardiac cava if the heart is not being retrieved) is incised to drain blood from the donor, and cold perfusion begins. Ventilation is stopped. Iced saline slush is poured over the organs to be recovered to provide additional surface cooling.


UW solution into superior mesenteric vein
Solution into infrarenal aorta
Effluent of blood from vena cava

88.5

When the organs are cold and blanched, they are removed. This requires care, as the blanched, non-pulsatile vessels are not so readily identified. The hepatic artery and celiac axis are removed en bloc with a patch or tube of aorta. The portal vein is retrieved with a stump of the splenic and superior mesenteric veins (or is incised at the level of the coronary vein if the pancreas is being retrieved). The right lobe of the liver and the vena cava are mobilized, retaining the right adrenal gland with the graft. The infrahepatic vena cava is transected just above the renal veins and the suprahepatic vena cava is divided with a cuff of diaphragm.

On the bench, the liver is further perfused with preservation solution via the portal vein. The bile duct is gently irrigated. The liver is packed in sterile plastic bags containing cold UW solution and stored in ice for transportation.

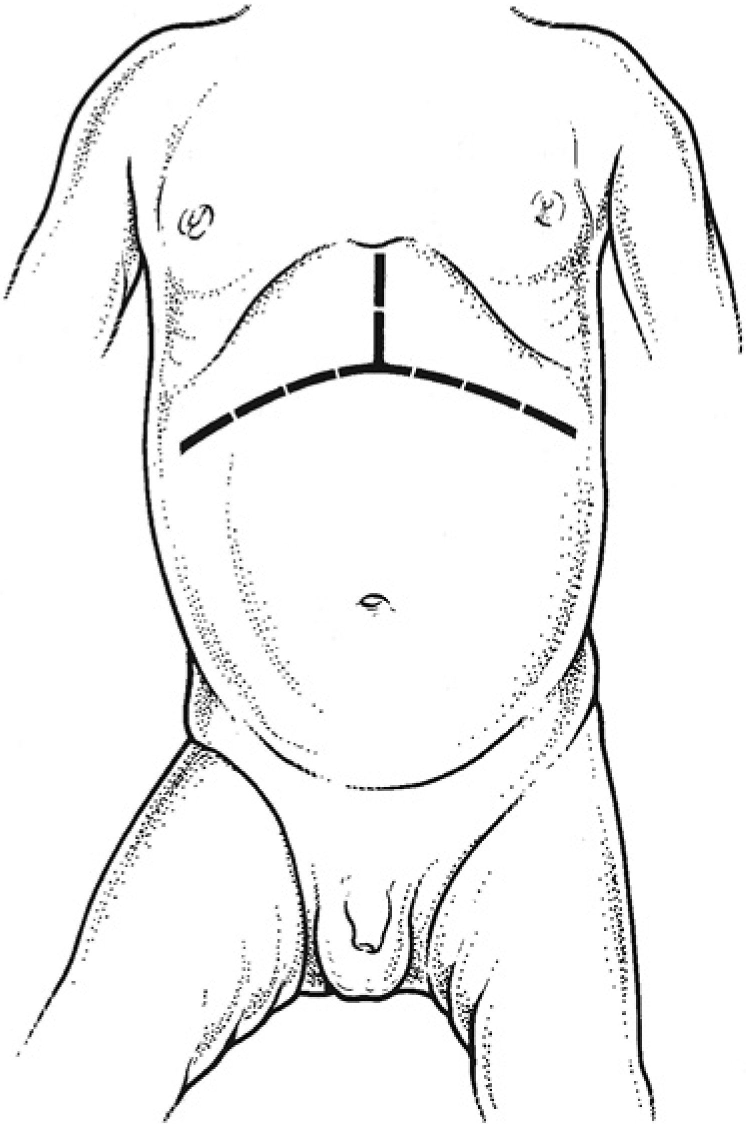
After the kidneys have been removed, the iliac arteries and veins are retrieved for any subsequent vascular reconstruction. Carotid arteries, jugular and subclavian veins may also be recovered to provide more options, given the anticipated size mismatch between the adult donor and the pediatric recipient. Back at the transplant institution, the liver is cleaned and prepared on the back table; redundant tissue is removed. The hepatic artery and portal vein are prepared for implantation.

RECIPIENT OPERATION

The recipient hepatectomy can be difficult in children with biliary atresia and portal hypertension in whom dense vascular adhesions from previous surgery may make dissection of the portal structures, duodenum, Roux loop, and transverse colon hazardous.

Incision

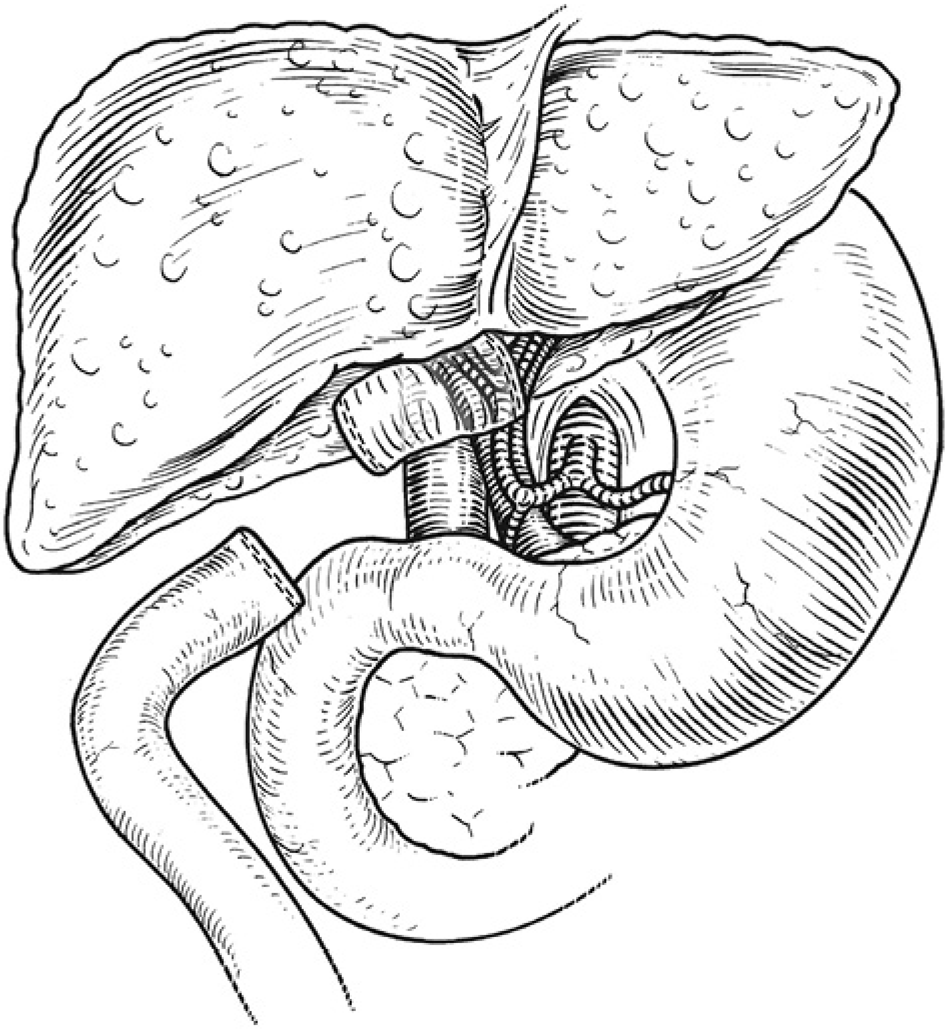
The abdomen is entered through a curved, bilateral subcostal incision and routinely extended vertically in the midline to the xiphoid process for better exposure, although it may be avoided in cases of small infants with a distended abdomen from ascites (**Figure 88.6**). The abdominal muscles are divided with cautery to maintain hemostasis. The liver is exposed using an upper abdominal retractor.



88.6

Hepatectomy

In children who have previously had a portoenterostomy for biliary atresia, the liver is separated from the abdominal wall with cautery. This dissection becomes easier once freer tissue planes are reached superiorly and posteriorly. The transverse colon and duodenum are carefully separated from the liver with the aid of precise bipolar cautery. The jejunal Roux loop is identified, divided, and oversewn in two layers close to the liver hilum in order to assure adequate length of the remaining Roux limb (**Figure 88.7**). The portal structures can then be dissected. In ALF, the porta hepatis is dissected before mobilizing the liver and the hepatic artery is ligated early to try to minimize hemodynamic instability.



88.7

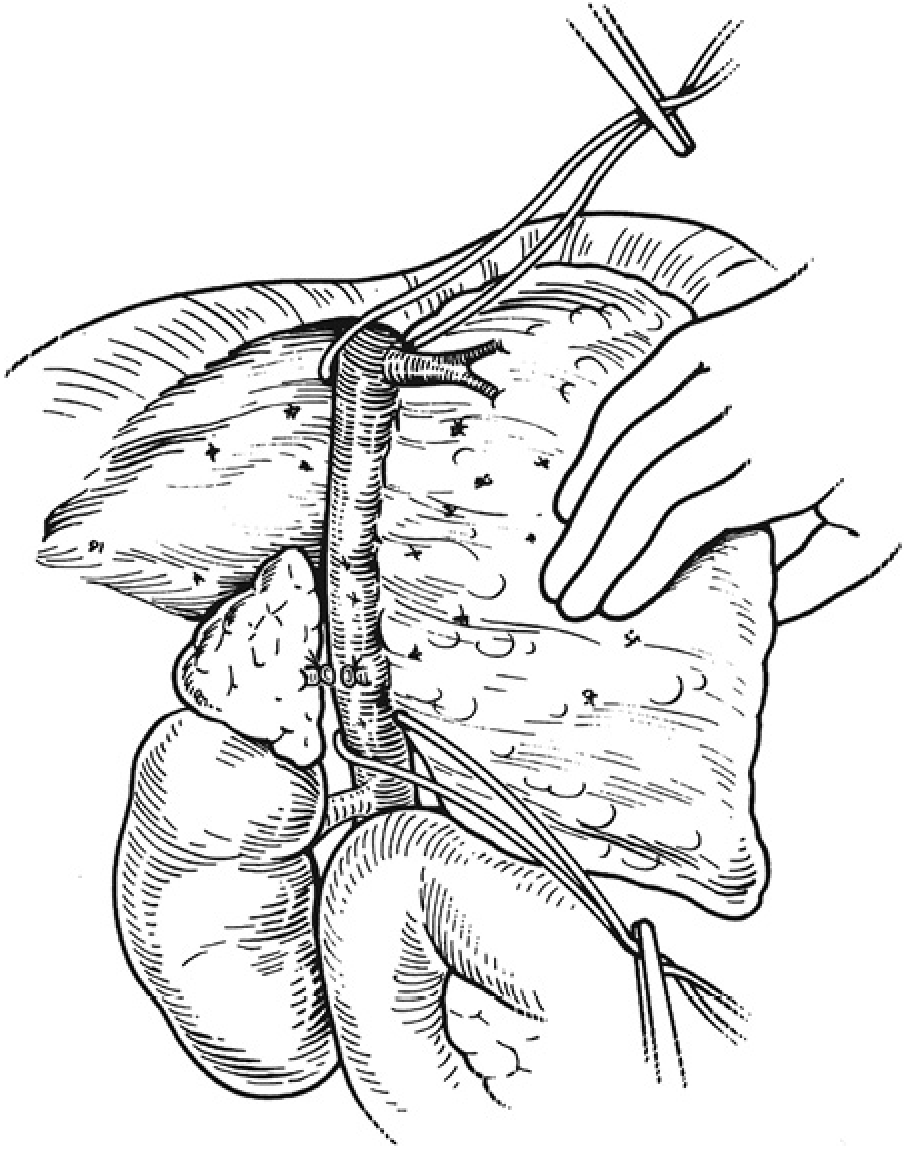
Where present, the CBD is divided between ligatures closer to the duodenum. The hepatic artery is traced to beyond its segmental branches, which are tied individually and divided. This is essential in order to maximize the number of candidate arteries available for possible use at the time of the arterial anastomosis. This constitutes a critical step in cases of living-donor intervention, as the donor artery is expected to be short in length and small in diameter. The portal vein is dissected and isolated; if the portal vein is narrow, either the bifurcation should be preserved or its origin exposed at the confluence of the splenic and superior mesenteric veins. Dividing the gastroduodenal artery helps to expose the proximal aspect of the portal vein and the confluence of the splenic and mesenteric veins.

If not previously divided, the falciform and triangular ligaments are incised. The gastrohepatic ligament is divided between ligatures. The liver is retracted medially and the right lobe is dissected from the diaphragm and retroperitoneum using cautery. The infrahepatic cava is encircled above the renal veins, and, using this plane as a guide, the right adrenal vein is identified and preserved or divided between ligatures if necessary. The plane behind the retrohepatic cava is carefully dissected from both sides until the suprahepatic vena cava can be encircled ready for subsequent clamping.

At this point, the dissection is completed and the diseased liver would be ready to be removed. Before proceeding, the anesthetist is consulted to ensure their readiness and also to verify the recipient's hemodynamic stability before going into the anhepatic phase.

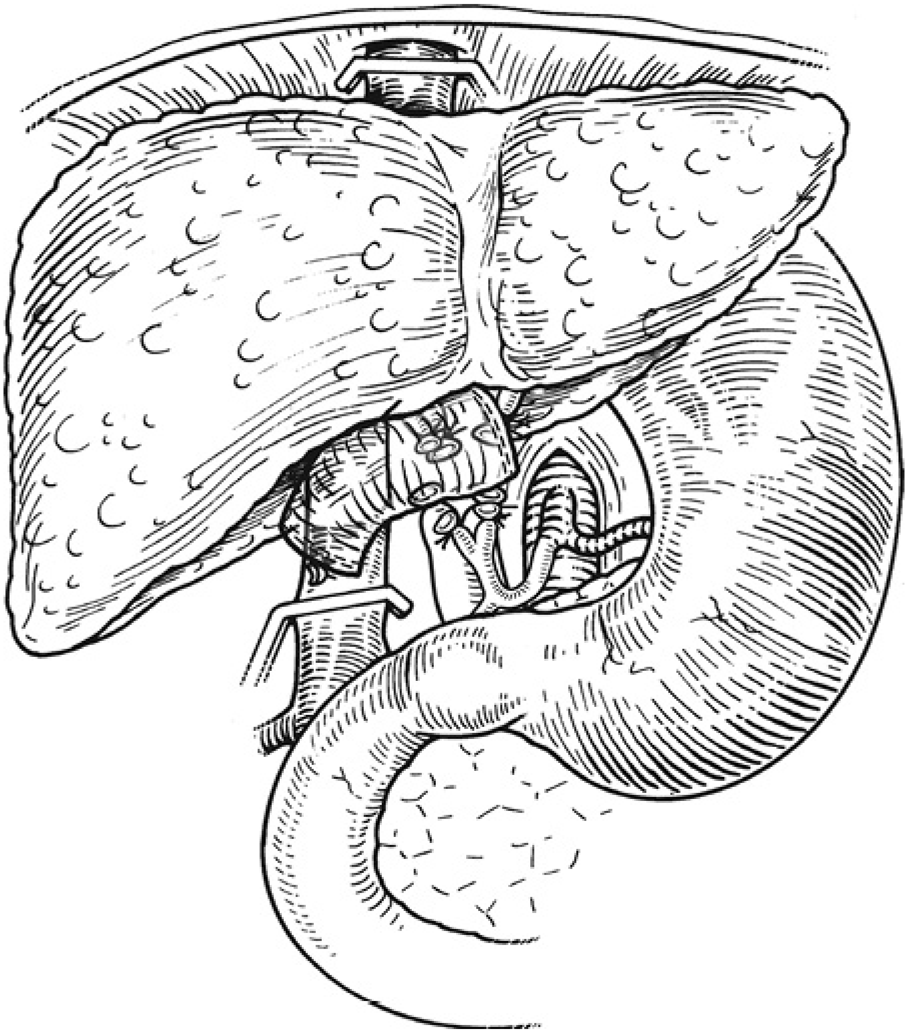
Some patients will require an aortic conduit, either because of their intrinsic disease (e.g. Alagille syndrome), or because of technical requirements (e.g. in deceased donor left lateral segment with dual arterial blood supply from a left hepatic artery and an accessory left hepatic artery coming off the left gastric artery, for which a common aortic Carrel patch will be used). The infrarenal aorta is preferred as an anastomotic site as it does not involve impairing arterial renal inflow during the anastomosis. The infra renal anastomotic site should be prepared before implantation of the new liver. A Cattell-Braasch maneuver is performed, reflecting the ascending colon and small bowel to the left of the abdomen, exposing the recipient's vena cava and aorta. The tissue overlying the aorta is incised, and lumber arteries often need to be ligated and divided to allow for aortic clamping. The inferior mesenteric artery may need to be divided, but most often can be preserved. A tunnel is created under the duodenum and over the vena cava through which the conduit will later be passed (**Figure 88.8**).

Preparing the infrarenal aorta beforehand is an important step in patients without portal hypertension, as once the liver is reperfused, evisceration of the small bowel will create a torsion effect on the portal vein inflow, and because of the lack of intra-abdominal varices, may result in low flow, venous congestion, and increase the risk of portal vein thrombosis.



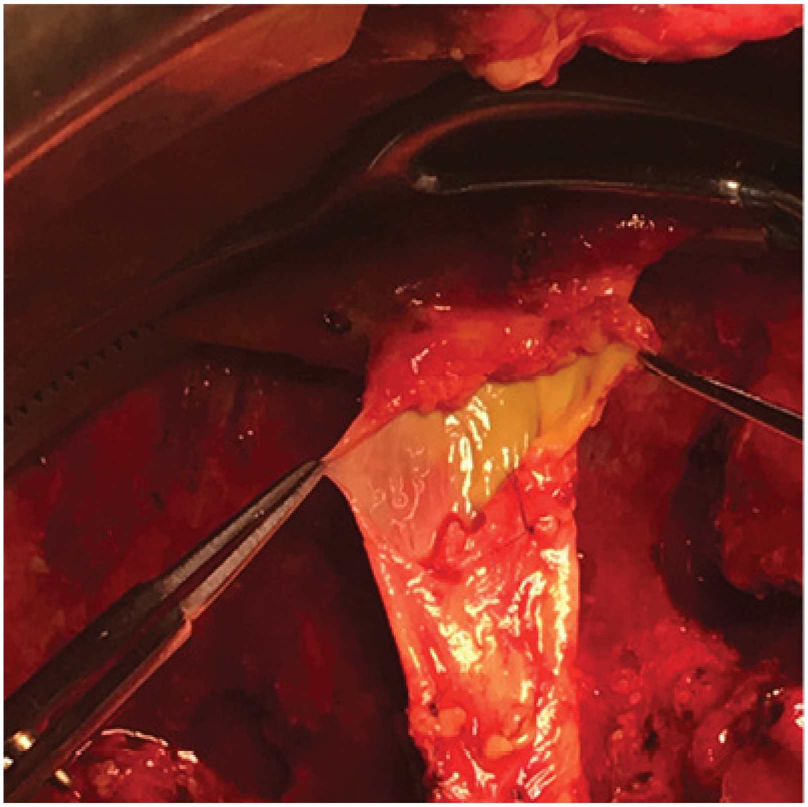
88.8

The portal vein is divided between ligatures or vascular clamps. Both the suprahepatic and infrahepatic vena cavae are occluded with secure vascular clamps; the former is clamped with a cuff of diaphragm, avoiding the phrenic nerve (**Figure 88.9**).



88.9

Provided the patient is hemodynamically stable, the liver is dissected sharply off the underlying vena cava. This technique is safer and less prone to bleeding than tying off all the retrohepatic tributaries in these often very hard and cirrhotic livers. Once the liver is mobilized off the inferior vena cava (IVC), the right hepatic vein is divided, and then the middle and left together. All venous tributaries on the retrohepatic cava are sutured or ligated. The right hepatic vein may be sutured or left open depending on the size of the liver to be implanted. For segmental 2–3 grafts or left lobe grafts (see subsequent sections), it may only be necessary to use the orifices of the left and middle hepatic veins opened together. For right lobe grafts, the right hepatic vein is left open and the left and middle are sutured shut. It is the authors' practice to also implant a whole liver in a "piggyback" fashion, for which all three hepatic veins are opened and joined together to form a venous cloaca to serve as the site of anastomosis to the donor (**Figure 88.10**).



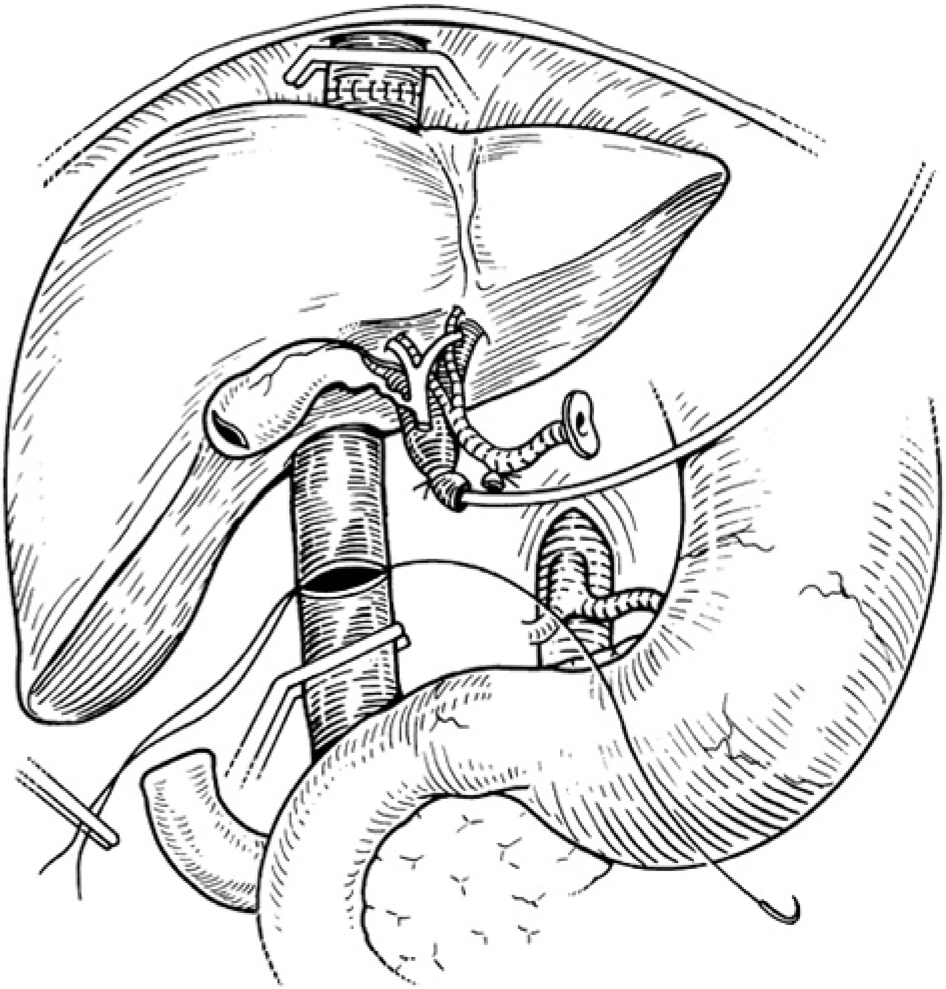
88.10 Hepatic vein orifices opened into a single orifice.

In cases where the recipient's vena cava needs to be resected, the suprahepatic vena cava is divided and extra length can be obtained by incising the liver at the confluence of the hepatic veins. The infrahepatic vena cava is then divided and the liver removed. Meticulous hemostasis of the retroperitoneum should be achieved with a combination of cautery, argon beam coagulation, and suture ligation. Access to this area is more difficult once the donor liver has been implanted.

Implantation

Liver implantation must be performed expeditiously to minimize the period of warm ischemia of the graft. Implantation begins with the suprahepatic vena caval anastomosis, which is performed with a continuous 5/0 PDS™ suture. In reduced donor graft, the hepatic vein should be kept short in order to avoid twisting on the venous pedicle. The vein must be widely anastomosed and properly oriented to the recipient cava to prevent venous outflow obstruction. The use of PDS™ absorbable monofilament suture is thought to prevent fixed anastomotic strictures that may ultimately prevent growth of the anastomosis as the child grows and slowly lead to an outflow obstruction. A double-ended suture is placed in each corner of the cava and the left side is tied as the liver is placed in the hepatic fossa. The posterior wall of the cava is anastomosed with a continuous everting suture, which ensures endothelial apposition. For transplants that do not involve a vena cava replacement, before completing the hepatic vein to vena cava anastomosis, residual preservative solution rich in potassium is flushed out of the liver with 500–1000 mL of cold lactated Ringer's solution infused via the portal vein, after which the anastomosis may be completed.

In cases of vena cava replacement, the infrahepatic vena cava anastomosis is performed next with an everting 5/0 PDS™ suture; the anterior wall is left incomplete and the liver is flushed of any residual preservative solution with an appropriate volume of lactate Ringer's solution infused via the portal vein. The effluent flows out of the incompletely anastomosed infrahepatic vena cava, which is then completed (**Figure 88.11**).



88.11

The portal vein anastomosis is performed next. Care must be taken to avoid redundancy or rotation that might cause kinking, but in cases of segment or lobe graft, the surgeon must allow enough vein on the recipient side in order to prevent undue tension in the anastomosis as the suture line falls more posteriorly as the liver is allowed to rotate back into position at the completion of the portal vein suture line. Before starting the anastomosis, the recipient portal vein should be flushed with heparinized saline solution to prevent clots from forming in the clamped vein. In children with no pre-existing coagulopathy (e.g. children with metabolic deficiencies such as urea-cycle defects) full heparinization is used since the prolonged clamping of the portal circulation may lead to mesenteric vein thrombosis or clots forming in the obstructed portal vein.

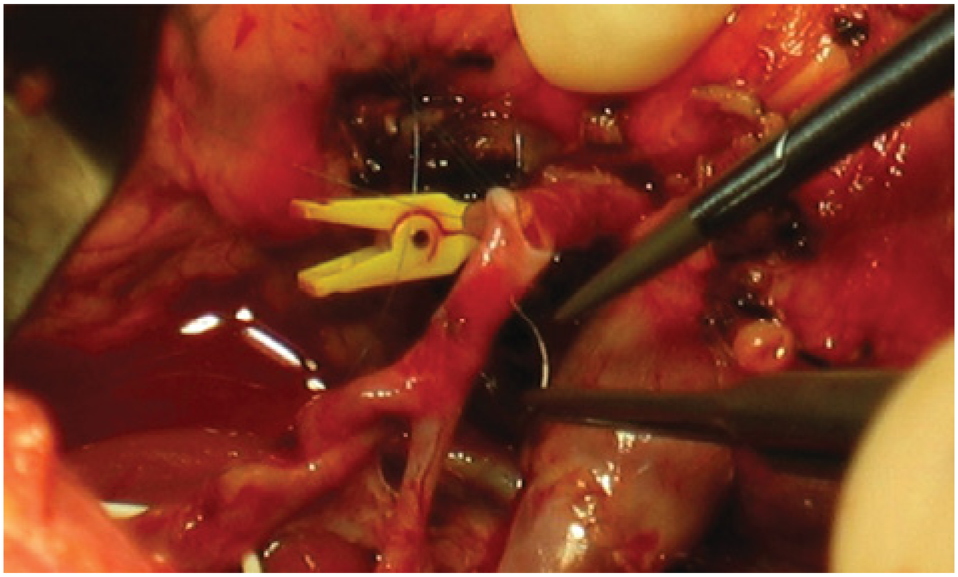
The anastomosis may be performed with continuous 6/0 or 7/0 polypropylene or PDS™ sutures. The authors prefer to perform the anastomosis using polypropylene, as they believe there is a lesser inflammatory reaction during suture reabsorption, which may help decrease the incidence of portal vein anastomotic stricture. Size discrepancies between the donor and recipient vein must be dealt with using a variety of potential techniques. These include "cheating" on the larger vein to compensate for the smaller circumference of the smaller vein, imbrication of the vein wall, spatulation of the smaller vein to increase the effective circumference, fashioning a branch patch from the bifurcation of the smaller recipient vein, or placing an interposition vein graft that is midway in size between the donor and recipient. At completion of the anastomosis, the knot is tied away from the vein wall ("air knot"), allowing the vein wall to expand freely with the influx of blood.

The anesthetist checks the patient for stability. In cases of lobe or segment transplant, before proceeding with vascular clamps removal, the cut surface is sprayed with fibrin glue to ensure further hemostasis. Then the suprahepatic caval clamp is removed first, followed by the infrahepatic caval clamp. A quick search is made for major bleeding points and the stability of the patient is confirmed before the portal clamp is removed and the liver reperfused. Any major bleeding points are identified and controlled. If still present, and in cases of whole liver graft, the gall bladder is removed using cautery and the cystic duct is transfixed and ligated with an absorbable suture.

The donor hepatic artery is gently flushed with heparinized saline and a soft bulldog clamp is applied to prevent backflow of blood from the donor liver, which would interfere with the arterial anastomosis. The recipient artery is also prepared by dissecting out toward the celiac axis, and freeing the branches from the surrounding connective, lymphatic, and ganglionic tissue for at least 1 cm beyond the take-off of the gastroduodenal artery. The surgeon must choose where to make the anastomosis based on the sizes of the various branches, and whether or not a conduit to the aorta is preferable to using the native hepatic artery.

The hepatic artery is anastomosed end-to-end or end-to-side depending on the anatomy and size discrepancies between the recipient and donor artery to either a branch patch of the recipient hepatic artery bifurcation or gastroduodenal artery. In whole liver transplants performed in larger recipients (teenagers) with both large hepatic arteries, the arterial anastomosis can be safely performed with a continuous 7/0 or 6/0 polypropylene suture posteriorly and interrupted sutures anteriorly. Magnification (with loupes or a microscope) is essential, even more so with small arteries. If the native artery is small or has poor flow, a donor iliac arterial conduit (or the celiac axis of the donor) anastomosed to the recipient's infrarenal aorta may be used to arterialize the graft.

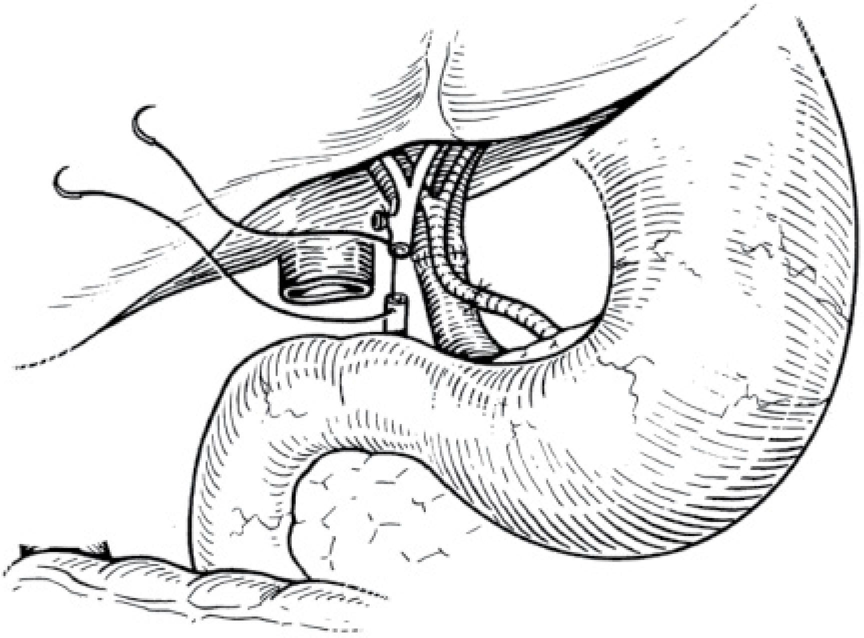
Interrupted 7/0 or 8/0 sutures are recommended in the smallest arterial reconstructions such as those in living-donor operations where the donor artery is usually either the left hepatic artery or the segment 2–3 artery and is usually 3–5 mm in diameter. This should be done with either 4.5× loupe or operating microscope magnification. The yellow plastic bulldog is used to gently occlude the recipient hepatic artery that is transected transversely. The anterior wall of the anastomosis is done with interrupted 7/0 or 8/0 non-absorbable monofilament sutures (**Figure 88.12**).

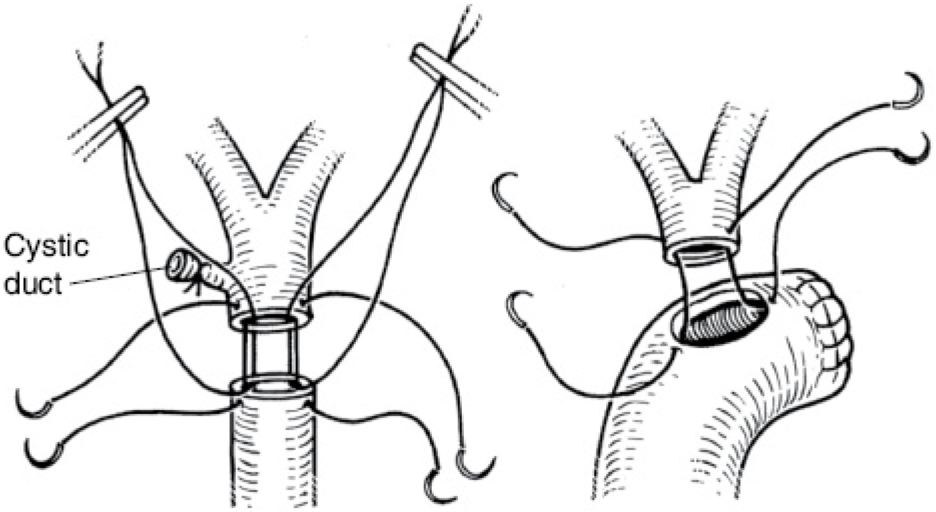


88.12 Hepatic artery anastomosis.

Before proceeding with the biliary anastomosis, verification of appropriate blood flow is performed. The use of adjuncts including intra operative Doppler, US, and flow probes will help in confirming clinical findings of recipient hemodynamic stability, but also color and appropriate fullness of the graft.

In most larger recipients (teenagers), and in cases of whole liver grafts, a duct-to-duct biliary anastomosis is performed using interrupted 6/0 PDS™ sutures (**Figure 88.13**). The donor duct may require trimming to produce a healthy vascular margin. The posterior row of sutures is placed before the ends of the ducts are approximated and the sutures tied.





88.13

A Roux-en-Y hepaticojejunostomy is performed in cases of biliary atresia and sclerosing cholangitis or if the recipient CBD is very small. Generally, duct-to-duct anastomoses are not recommended in children <10 kg in weight or in those who receive a split-liver transplant even if the entire duct accompanies the graft.

TECHNICAL VARIANT GRAFTS

Most deceased and all living donors are adults. As a result of improved pediatric medical care, size-compatible donors for infants or small children who are on organ waiting lists are rarely available. In the current situation of organ/donor shortage, reduction techniques have been developed to make it possible for pediatric liver recipients to benefit from adult organ donation.

Segmental liver grafts are prepared from whole deceased donor livers and from living donors. In practice, there are three main types of reduced-size graft:

- segments 2–3 grafts
- Left lobe (segments 2–4)
- Right lobe (segments 5–8)
- monosegmental grafts (segment 2)

The type of graft is determined by the relative sizes of the donor and recipient and the individual anatomical characteristics of the donor. With cadaveric organs, the whole liver may be split so that two patients may be transplanted with a single organ. Reduced-size transplantation implies that an organ is reduced in size to accommodate a liver too large to be transplanted whole, but that only one patient is transplanted. Reasons for not splitting but simply reducing an organ in size may be anatomical, surgeon preference, or that the liver is too small to adequately serve the needs of two individuals. Split-liver grafts are preferable to reduced grafts because this maximizes the donor organ pool.

Donor criteria for split or reduced grafts are more stringent than for whole grafts because of the extra insult of the bench procedure: Donors under age 50 years, less than 10% macrovesicular fat, minimal pressor support, near-normal liver enzymes and bilirubin, and less than 5 days in hospital at the time of the procurement of the organ. Split-liver transplant may be performed *in vivo* at the time of the donor operation, or *ex vivo* once the recovered organ is brought back to the primary recipient transplant center. While *in vivo* split-liver transplant has the advantage of reducing cold ischemic time, it does increase significantly the duration of the donor operation, putting the patient at risk for intraoperative complications, with the possible theoretical risk of intraoperative donor death, and therefore loss of all allocated organs. The *ex vivo* "back table" split-liver technique does increase cold ischemia time of the liver, but it allows for a standard quicker and safer donor surgery. It also allows for excellent anatomic visualization of the liver, including variants in segment 4 arterial supply and bile duct anatomy.

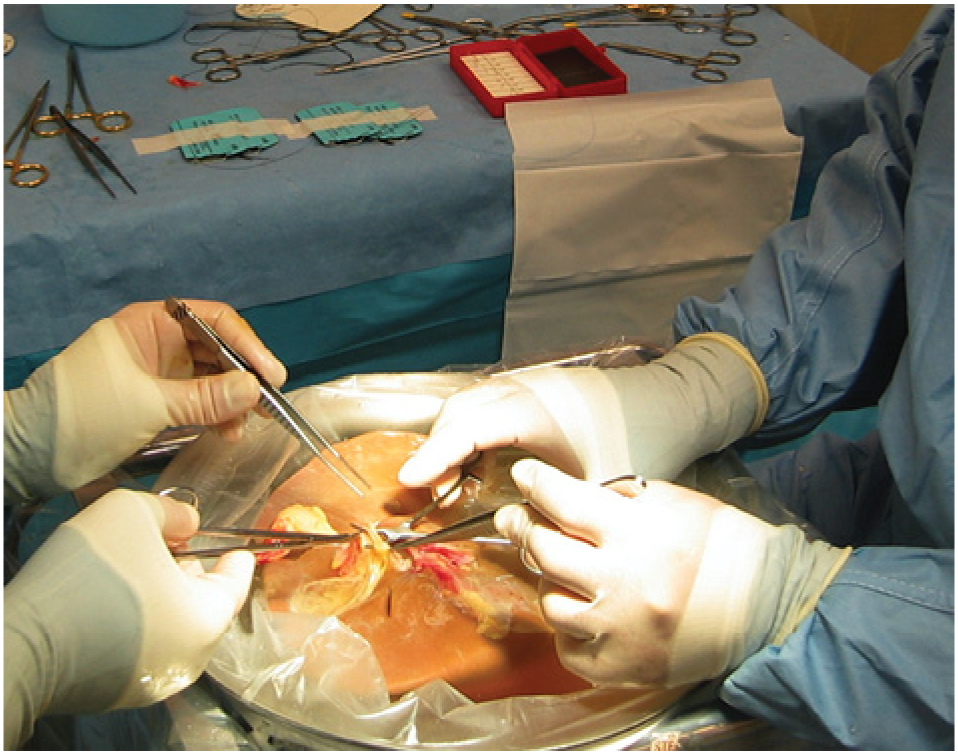
Various size guidelines are helpful in pediatric LT. As a minimum, the graft weight should be about 0.8% of the recipient's body weight in living-related LT; a greater safety margin is advisable in deceased donor transplantation, especially in small infant recipients. The adult liver usually weighs between 1000 g and 1700 g. The left lateral segment comprises about 25% of the liver weight and the left lobe about 40%. The maximum graft size that can be safely accommodated in the child's abdomen depends on the recipient's weight, the presence of ascites and hepatomegaly, and other factors, but, as a rough guide, a donor-to-recipient weight ratio of up to 1.5 : 1 is often acceptable for whole livers and right lobe grafts, up to 4 : 1 for a left lobe graft, and up to 13 : 1 for a left lateral segment graft. However, because of the exceptionally wide variation in the relative size of the segment 2–3 portion of an adult liver, it must always be necessary to visually inspect the organ prior to deciding on the anatomical unit appropriate for any individual recipient.

Small-for-size syndrome is defined as by a graft-to-recipient weight ratio of <0.8. It is associated with severe ascites, postoperative prolonged hyperbilirubinemia, coagulopathy, and possible encephalopathy. While it is not encountered as often in the pediatric population, it remains a consideration and possible complication in living-donor LT, as preoperative imaging and segment volume estimation may actually be inaccurate and underestimated.

The use of technical variant transplants has greatly reduced waiting list mortality because most donors are adults and most pediatric recipients are infants or small children. Still, mortality in very small children requiring small segmental grafts remains high. Monosegmental transplantation has been used to further reduce the size of a segment 2–3 graft to allow even greater discrepancies in the donor-to-recipient weight ratio.

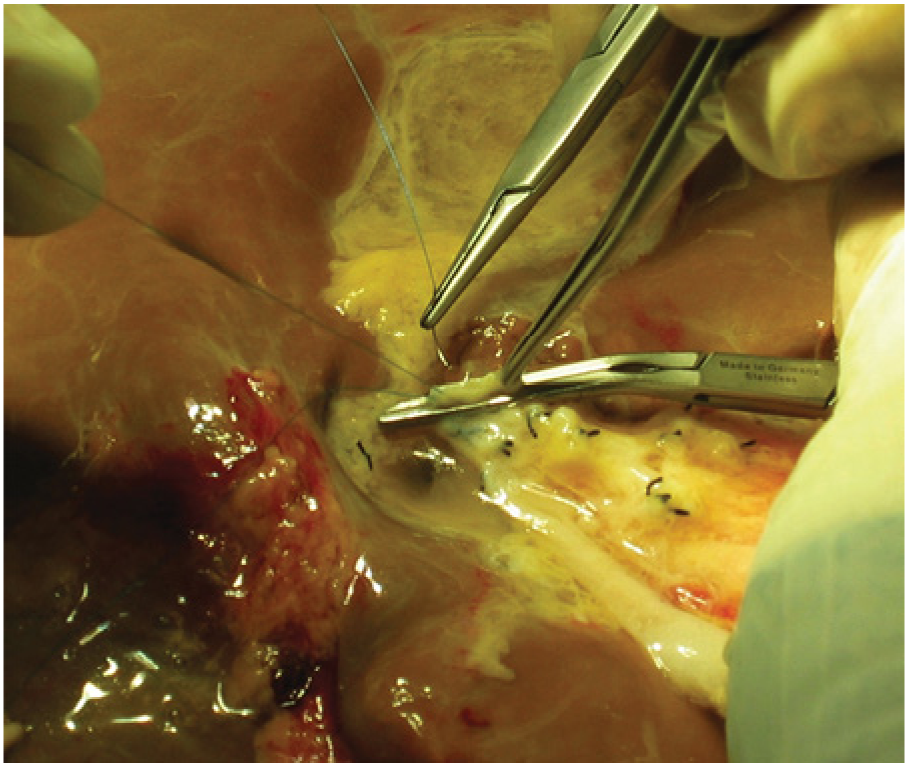
Reduced grafts

Liver reduction is best performed as a bench procedure in the recipient operating suite (**Figure 88.14**). The donor liver must be kept totally immersed in cold UW solution at 4 °C during the procedure. Care must be taken to avoid excessive traction on the hepatic artery, which could cause intimal injury.



88.14 Back table liver preparation.

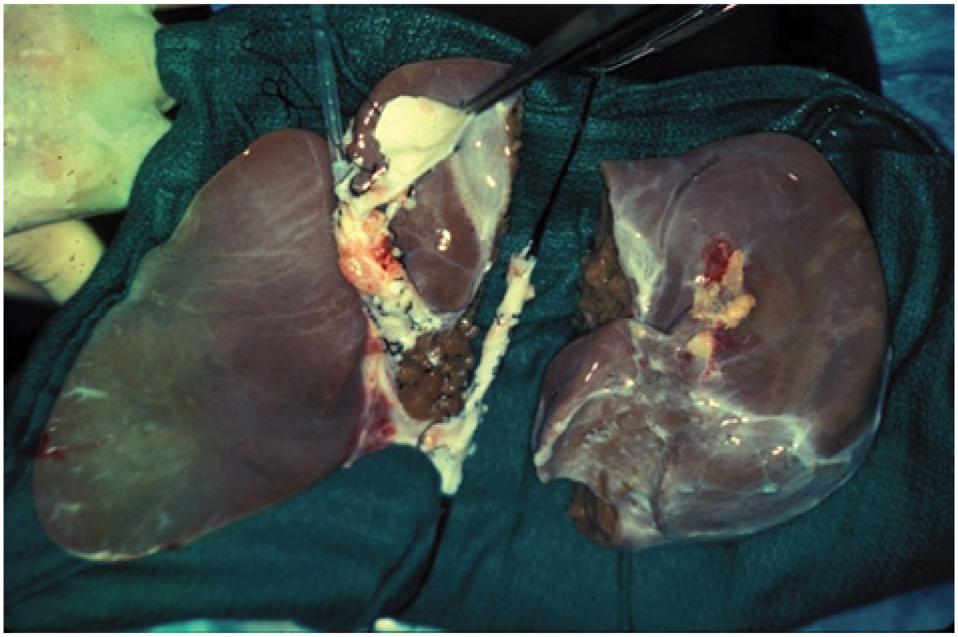
With right or left lobe grafts, the parenchyma is transected alongside the principal plane slightly to the left or right of the middle hepatic vein, respectively. The interlobar plane is defined by the gall bladder fossa anteriorly and the bed of the IVC posteriorly for a reduced-size transplant. In **Figure 88.15**, the right portal vein has been transected next to the gall bladder fossa and is being oversewn in preparation for transplanting only the left lobe.



88.15 Oversewing the right portal vein stump on the back table.

Defining the venous drainage is critical. The middle hepatic vein accompanies either the right or left lobe along with either the right or left hepatic vein. The main branches to the lobe that is being discarded are ligated well away from the midline, but outside the parenchyma, so that they are effectively ligated and defined. If the recipient's IVC needs to be resected en bloc with the liver (sometimes required in cases of transplant for liver tumor with encasement or invasion of the vena cava), it is preserved with the graft. Otherwise, the vena cava is excised in order to solely preserve a common hepatic vein ostium, since most reduced grafts are implanted using the piggyback technique. If the left/right and middle hepatic vein ostia insert separately in the vena cava or if they are too far away from one another, they may need to be reconstructed as a single ostium to simplify further anastomosis. Bile duct radicles and portal vein branches to the caudate lobe are also divided since the caudate lobe is rarely used in reduced size transplants.

Figure 88.16 shows that the liver has been divided into right and left lobes, and the left lobe (on the left) has the IVC and the aorta attached to the hepatic veins and hepatic arteries.

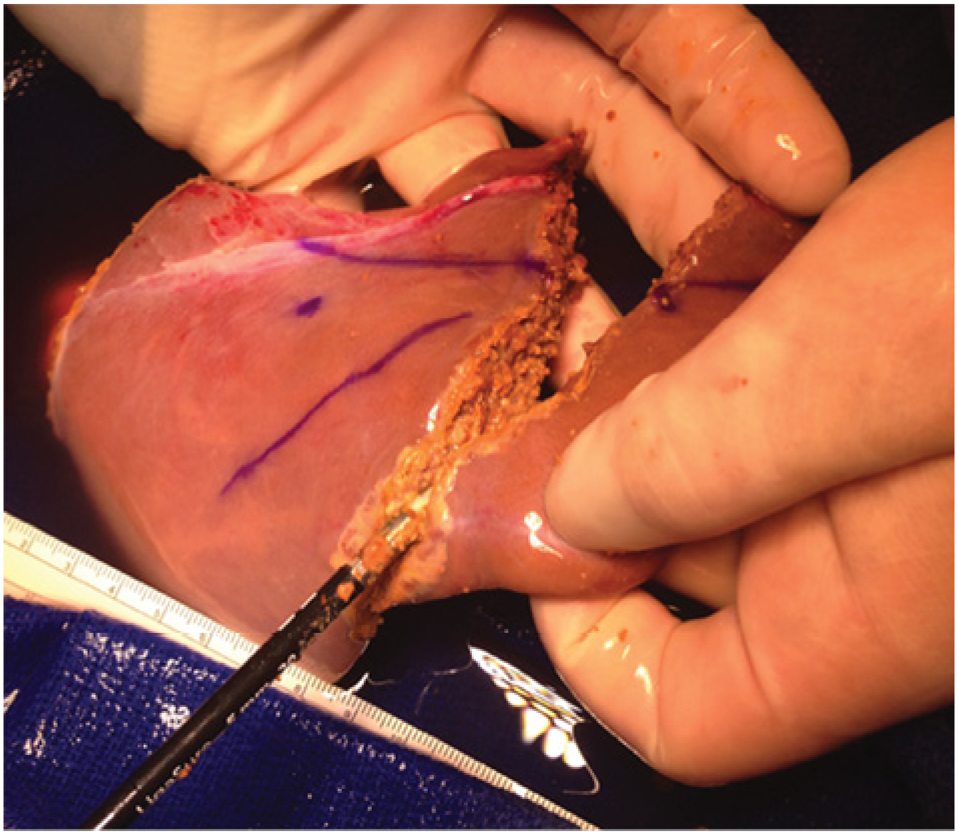


88.16 Right and left lobes of the liver.

Biliary and vascular branches on the cut surface of the graft are carefully ligated. LigaSure™ (Valleylab, Boulder, CO, USA) instrumentation has replaced the finger fracture technique or manual ligating of most biliary or vascular structures crossing the plane of transection. After division, the cut surface is tested for leaks by gently perfusing preservation solution through the portal vein. These are oversewn. After implantation, and prior to graft reperfusion, the cut surface will be sprayed with fibrin glue to ensure further hemostasis. The reduced graft comes with the main portal vein, the celiac axis, the main bile duct, and possibly the IVC, but most of these structures may be removed and only the branches to the lobe may be used at the time of implantation. In general, preserving the complete vascular structures and using them is unnecessary.

In preparing a left lateral segment graft, the hilar dissection is very similar to that used for a left lobe graft except that the plane of parenchymal transection is just to the right of the falciform ligament. The right-sided branches of the portal triad are dissected and divided outside the liver; the stump of the right hepatic artery is ligated, and the right branch of the portal vein is closed with a continuous polypropylene suture. Portal vein branches to segment 4 are divided within the Rex recessus. Caudate branches from the origin of the left portal vein are divided. The left hepatic vein is incised close to the liver parenchyma, unless vena cava replacement is required. In this case, the middle and right hepatic veins would be divided from the vena cava and their orifices oversewn. The common hepatic artery and main portal trunk are retained with the graft. The main bile duct is preserved in continuity with the left hepatic duct. Care must be taken not to injure the left hepatic duct at the base of segment 4, so the parenchymal division should be away from the hilum as it nears the hilar structures.

Afterwards, the surgeon may elect to cut back the duct, and make the biliary anastomosis at the level of the common hepatic duct or even the left hepatic duct. For LT in small infants, the left lateral segment can be further reduced, creating a monosegmental graft. In the example shown in **Figure 88.17**, the paths of the left hepatic vein and the left portal vein have been drawn on the surface of the segment 2–3 graft and serve as a guide to the further reduction in mass to result in a monosegmental graft. Segment 3 is resected with the LigaSure™ (Valleylab, Boulder, CO, USA), preserving the hepatic and the portal veins, leaving segment 2 to be transplanted. The bile duct anatomy at the cut surface is not affected.



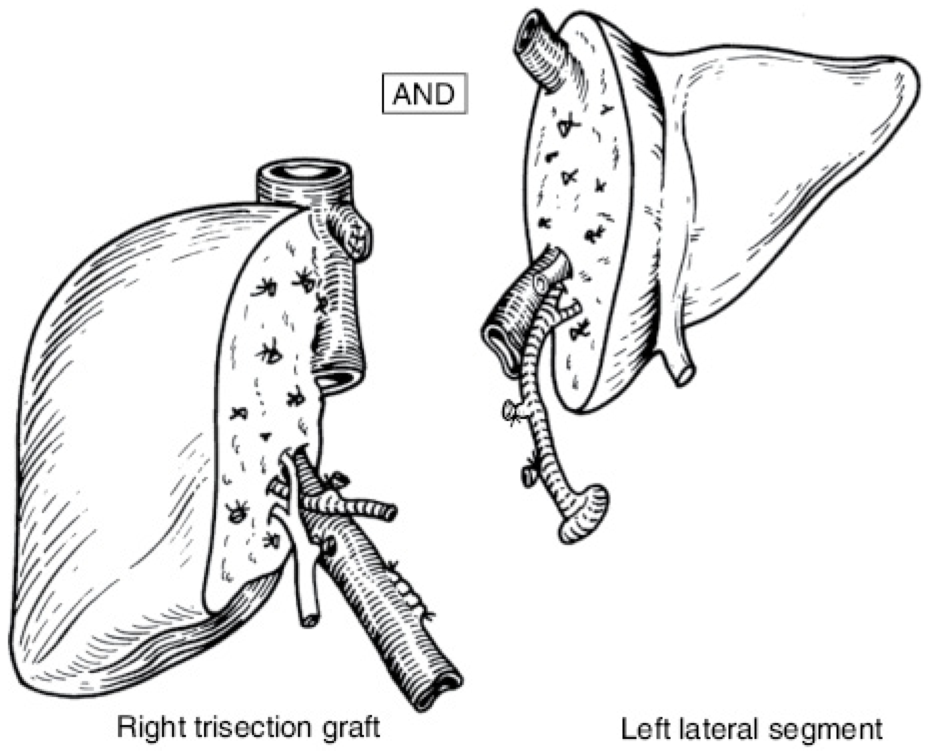
88.17 A monosegmental graft.

Split-liver grafts

The donor liver is divided into two grafts for transplantation into two recipients. The procedure is more complex than a size reduction since biliary drainage, venous drainage, and portal and arterial supply to both portions of the liver must be preserved intact.

Early assessment of vascular and biliary anatomy is important to ensure that there is no anatomic contraindication to splitting. The gall bladder is removed and the portal vein and hepatic artery carefully dissected to their bifurcations. Arterial anomalies often complicate the split procedure.

Left lobe–right lobe splits are less common than segments 2–3/right trisegment grafts, where the right trisegment goes to an adult or a teenager, and the segment 2–3 graft to a baby or small child (**Figure 88.18**).



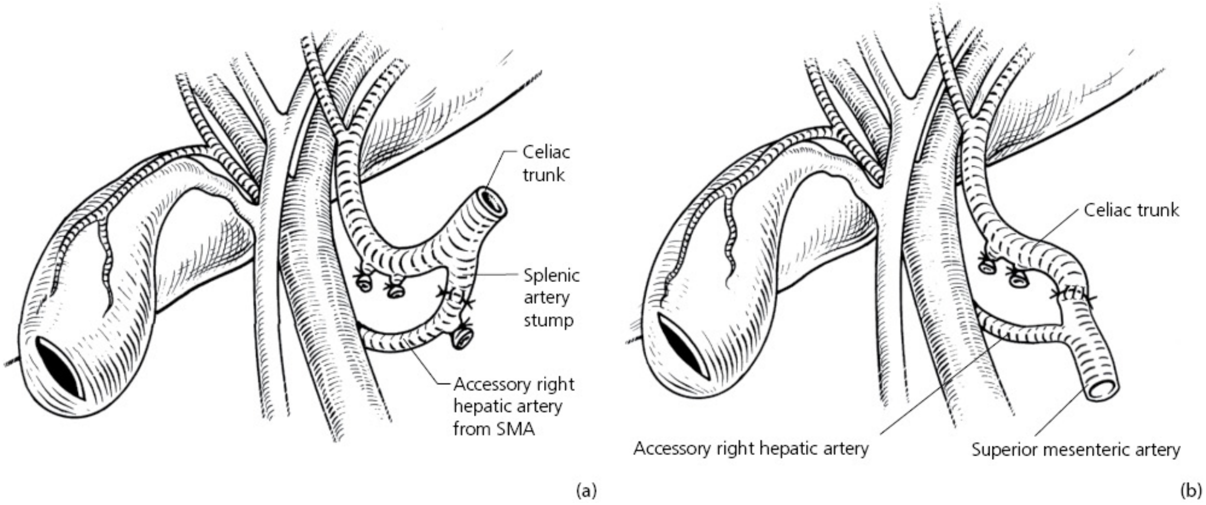
88.18

For the segment 2–3/right trisegment grafts split, it is important to try to preserve the arterial supply to segment 4, since the portal blood to segment 4 will always be divided. Strict allocation rules have been developed, so that allocation of vessels will be determined primarily by the center to which the primary offer has been made.

If segment 4 artery comes off the left hepatic artery, that vessel may be sacrificed, rendering segment 4 ischemic. The right trisegment graft may still be usable since intraparenchymal collaterals may exist, but the center using the right trisegment graft should realize the risk. The middle hepatic vein goes with the segment 2–3 graft, as does the bile duct, vena cava (if needed), and main portal vein. The celiac axis can go to the primary recipient if a child and then the right trisegment graft will just have the right hepatic artery. When the adult is receiving the primary offer, the child will generally be allocated the left hepatic artery with or without the segment 4 vessel. The parenchymal transection is along the same plane as for reduced-size livers.

Less commonly, right lobe–left lobe splits are performed. Even though the parenchymal transection is thicker, the hilar dissection is simplified since the arterial supply is always determined by a left–right hepatic artery allocation, with the celiac axis usually going to the graft that has been allocated to the primary recipient. Arterial anomalies complicate any split procedure, and may preclude safe utilization of both pieces. An anomalous left hepatic artery from the left gastric may simplify the arterial division in a segment 2–3/right trisegment split, as will an anomalous right hepatic artery coming from the superior mesenteric artery.

Arterial reconstruction on the back table may be required to simplify implantation of a liver segment with an anomalous hepatic arterial supply (**Figure 88.19a and b**). For example, a right accessory artery arising from the superior mesenteric artery (SMA) can be anastomosed to the splenic or gastroduodenal stump. An anomalous left artery arising from the left gastric artery requires no particular reconstruction since both the left gastric and common hepatic artery can be revascularized from a Carrel patch on the celiac axis.


Celiac trunk
Splenic artery stump
Accessory right hepatic artery from SMA
(a)
Celiac trunk
Accessory right hepatic artery
Superior mesenteric artery
(b)

88.19a,b

Extreme care must be taken in any split procedure in both donor selection and in careful technique during the split procedure since the lives of two recipients will depend on the adequate function of both halves of the split liver.

Living-related grafts

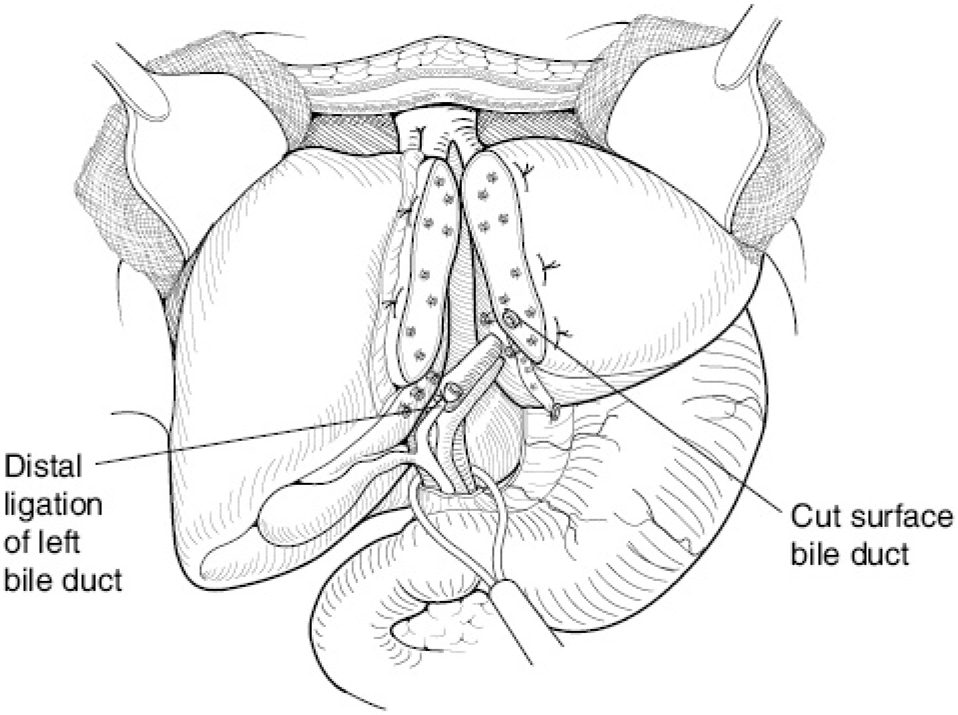
The greatest experience is with living-related donation of the left lateral segment from parent to child but, in recent years, living-related donation of the left lobe and adult-to-adult right lobe donation have become increasingly popular. Living-related donation has additional ethical and medical issues.

The safety of the donor is paramount. The potential donor should have a compatible blood group and appropriate liver anatomy. In the elective setting, the donor is evaluated for emotional stability and screened for any evidence that they may be subject to subtle coercion to donate. Emotionally related donors who do not have any direct family connection to the donor may also volunteer to be considered as donors when the parents or direct relatives are not suitable. Social media has become a novel way for families to reach out for possible living donors, but this may create an unexpected burden on donor centers, which need to screen multiple potential donor candidates who may not fully appreciate the concept of living-liver donation. In addition, altruistic individuals who step forward as donors for patients they do not know, so-called "Good Samaritan donors," must be carefully evaluated both medically and psychologically before being accepted.

Although the live-donor liver operation was first conceived as an elective procedure with adequate time for the prospective donor to come to a full realization of the risks involved, more recent experience has included live-donor transplantation for urgent situations, such as fulminant liver failure or acute decompensation of children with chronic liver disease. The familiarity of the public with living-donor transplantation has grown to the point where many parents demand to be considered as donors, even in the setting of fulminant failure, if they think that donating will increase their child's chances of surviving.

The donor operation is started first. When direct visual confirmation of hepatic suitability is obtained, the recipient operation is started. The authors advocate that the adult operation takes place in an adult-care institution by a team of surgeons separate from the one that will be caring for the child. For a left lateral segmentectomy, an upper midline abdominal incision is used to mobilize the segment 2–3 graft. The left side of the liver is mobilized by dividing the triangular ligament and the gastroduodenal ligament. The left hepatic vein and left portal triad structures are isolated with slings. The parenchyma is transected at the falciform ligament using LigaSure™ (Valleylab, Boulder, CO, USA) instrumentation.

When the parenchymal division is almost complete, the left portal vein is mobilized (dividing caudate branches) and the hilar plate and left bile duct divided (**Figure 88.20**). The segments 2 and 3 ducts may be separate at the line of transection.



88.20

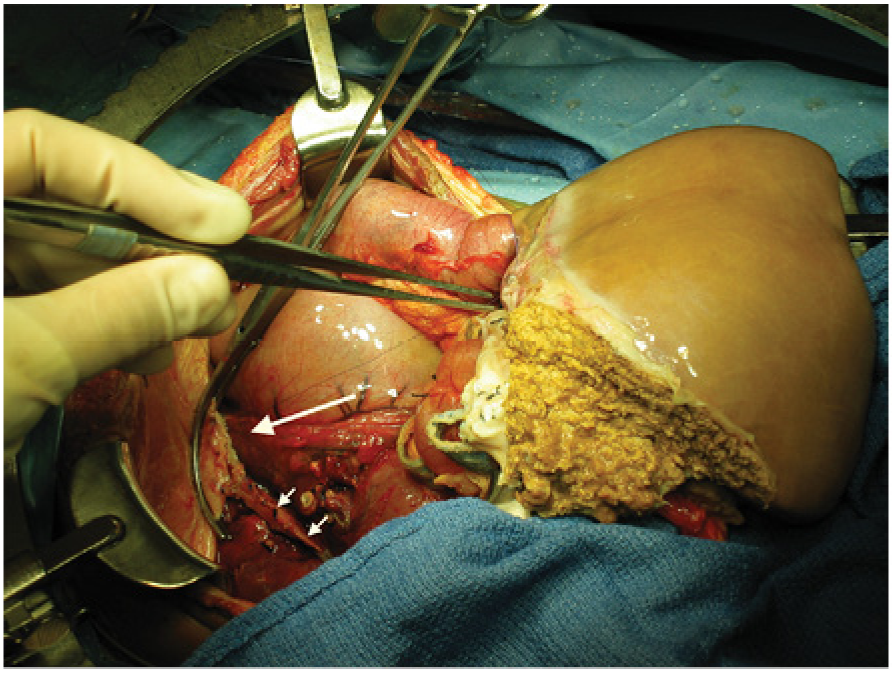
After completion of the parenchymal section, the left portal vein, hepatic vein, and hepatic artery are divided and the liver graft is removed and perfused with ice-cold UW solution on the back table.

For left or right lobe grafts, the liver is mobilized laparoscopically, and much of the parenchymal transection is also done laparoscopically with LigaSure™ (Valleylab, Boulder, CO, USA). The final division of the vessels is done through a midline incision, and the graft removed.

Laparoscopic mobilization has benefited donors in that the need for a large subcostal incision on both sides of the abdomen has been eliminated and the postoperative recovery is much faster and less painful.

Technical graft implantation

Figure 88.21 shows the left lobe graft of an older pediatric deceased donor being implanted into a child with fulminant failure. The orifice of the confluence of the left and middle hepatic veins is seen underneath the suprahepatic vena cava clamp (long arrow). This liver is being piggybacked onto the recipient cava, which is seen as a thin collapsed structure running down from the vena cava clamp (short arrows).



88.21 Implantation of a segment 2–3 graft.

A segment 2–3 graft implantation varies according to whether the graft originates from a live donor or is a reduced graft from a deceased donor. The technique used for a split-liver graft implant is similar to that utilized for a live-donor organ. The main difference is in the length of vessels available that would permit reconstructive procedures if necessary. In a live donor or split graft, the length of the portal vein and hepatic artery is quite limited unless the celiac axis is donated with the deceased donor split graft.

Even with a reduced donor graft, the left hepatic vein should be kept short in order to avoid twisting on the venous pedicle. The donor vein is anastomosed to the confluence of the left and middle hepatic veins of the recipient and that opening may be further extended onto the ventral surface of the IVC. The vein must be widely anastomosed and properly oriented to the recipient cava to prevent venous outflow obstruction. The anastomosis is carried out with PDS™ sutures. The absorbable suture is thought to prevent fixed anastomotic strictures that may ultimately prevent growth of the anastomosis as the child grows and slowly lead to an outflow obstruction.

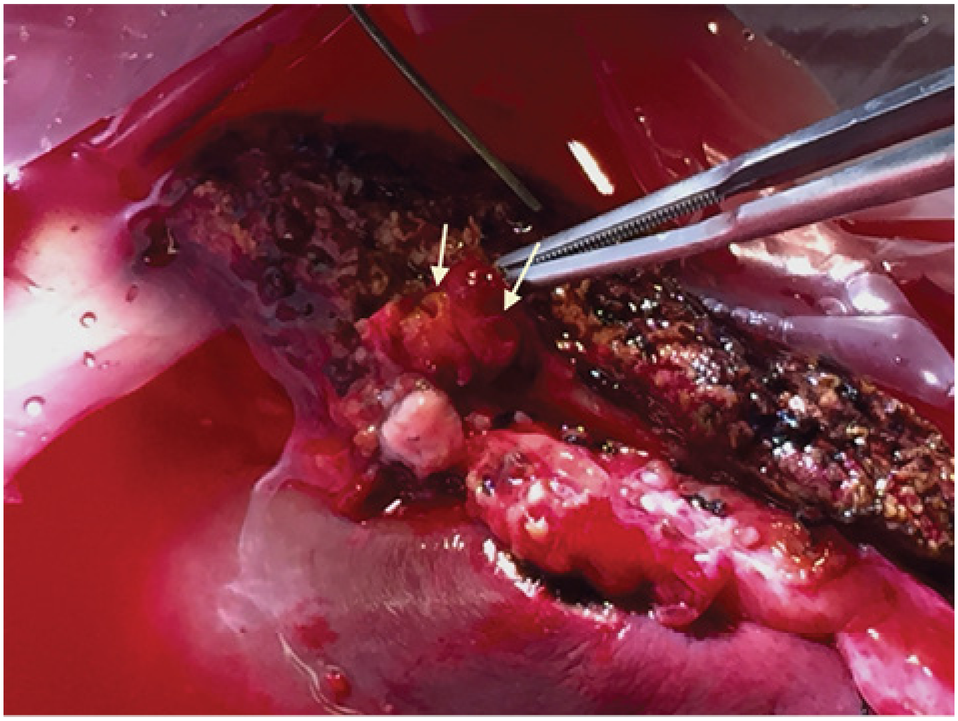
The anastomosis is started by placing two corner sutures, tying the left superior one, and proceeding by anastomosing the back wall from inside the vein and progressing to the right inferior corner. The front wall is done next from left to right and the liver is flushed through the portal vein with 500 mL of cold lactated Ringer's solution prior to completion of the hepatic vein anastomosis.

The portal vein is done next. Great care must be taken to allow enough vein on the recipient side to prevent undue tension in the anastomosis as the suture line falls more posteriorly as the liver is allowed to rotate back into position at the completion of the portal vein suture line. Different techniques (e.g. donor venous conduits) have evolved to compensate for the size discrepancies often observed between the portal veins of the donors and the often atretic or hypoplastic portal veins of the recipients.

For living-donor and split-liver transplants when only a short segment of donor left hepatic artery is available, the hepatic artery anastomosis is done to the recipient vessel at a location that more closely approximates the diameter of the donor artery. When the celiac axis is available, the surgeon may choose to anastomose the entire artery with a Carrel patch directly to the infrarenal aorta of the recipient. Alternatively, a donor iliac artery interposition graft may be placed on the recipient aorta and anastomosed to the celiac axis when the length of the celiac axis will not permit direct aortic anastomosis. Interrupted 7/0 or 8/0 sutures are recommended in the smallest arterial reconstructions with 4.5× loupe or operating microscope magnification in living-donor and split-liver grafts, as the arteries are frequently no more than 2–3 mm in diameter.

The 40 cm retrocolic Roux loop is anastomosed to the bile duct(s) at the cut surface using interrupted 6/0 PDS™ sutures.

Separate segmental ducts are seen in one-half to one-third of segment 2–3 transplants (**Figure 88.22**). In these cases, there have to be two separate biliary anastomoses. In reduced-size grafts, the anastomosis can be anywhere from the CBD to the left hepatic duct, but it is advisable to shorten the duct as much as seems convenient in order to reduce the chances of ischemic strictures from developing. The use of biliary stents is favored in some centers and may be helpful in facilitating biliary drainage in the early postoperative period when there may be bile duct anastomosis edema. However, the authors use a biliary stent when performing small biliary anastomosis mostly to avoid inadvertent closing of the bile duct when doing the anterior aspect of the anastomosis by including part of the posterior wall.



88.22 There are often two ducts (white arrows) in split and live-donor grafts, one each from segments 2 and 3.

Suturing the remnants of donor and recipient falciform ligaments helps to stabilize the graft and reduce the risk of torsion of the left hepatic vein. Care must be taken to close all the mesenteric defects created by the confection of the Roux loop, in order to prevent future internal hernias from occurring.

Hemostasis and closure

Complete hemostasis is secured. All anastomoses are checked. Bleeding from the raw cut surface of the segmental graft is controlled by a combination of fibrin glue and other topical hemostatic agents, superficial sutures, and argon beam coagulation. If bleeding is severe or persistent, venous outflow obstruction must be excluded. Otherwise, bleeding at this stage is often secondary to coagulopathy, and thromboelastography may be useful in guiding the use of blood products. Before proceeding with abdominal closure, suturing the remnants of donor and recipient falciform ligaments helps to stabilize the graft and reduce the risk of torsion of the left hepatic vein. The abdominal wall is closed in one layer in an interrupted fashion using absorbable suture, after placing two soft multi-fenestrated large-bore (10 Fr) silicone drains posteriorly in the right upper quadrant by the cut section in cases of a segment/lobe graft, and/or under the porta hepatis when a whole liver graft is used, and a duct-to-duct anastomosis is performed. The skin is closed with metallic staples. A prosthetic patch (e.g. Vicryl™) or skin closure alone (with delayed muscle closure) may occasionally be necessary to avoid an excessively tight abdominal closure.

POSTOPERATIVE CARE

Prostaglandin E infusion as a continuous drip is started immediately after the operation in all split- and reduced-liver transplants at a dose of 0.01 μg/kg per minute and increased hourly to a maximum of 0.05 μg/kg per minute as long as the blood pressure is maintained. This decreases ischemic–reperfusion injury and prevents primary non-function of the graft. It is almost never necessary in living-donor grafts since the preservation time is quite short.

Hemoglobin levels are maintained at 8–10 g/dL. During the first week, abdominal Doppler US scans help to identify any vascular or biliary complications. CT imaging provides better definition of problems identified with US screening.

Postoperative chest radiographs may show a right-sided pleural effusion, but this typically resolves spontaneously. Paresis of the right hemidiaphragm from a clamp injury to the phrenic nerve may cause prolonged dependence on ventilatory support, but only rarely is a diaphragmatic plication necessary.

Prothrombin time, INR, blood gases, plasma lactate, glucose, and electrolytes are checked regularly and serve as an index of early graft function. Nasogastric drainage is discontinued as soon as the ileus resolves and enteral feeding is started, usually within 48–72 hours. An acid suppressant is given routinely.

A detailed discussion of immunomodulatory regimens is beyond the scope of this chapter, but the overall strategy remains fairly simple: Minimizing adverse effects of immune suppressants, namely bacterial, viral, or fungal infection and malignancy while optimizing graft survival by preventing rejection. There is no universally accepted standard regimen immunomodulation following pediatric LT. At the authors' center, a single preoperative dose of 10 mg/kg of methylprednisolone is administered. About 12 hours postoperatively, both mycophenolate (10 mg/kg/dose) and tacrolimus (0.1 mg/kg/dose) are given twice daily with a tacrolimus level desired range of 10–12 ng/mL. Methylprednisolone is tapered over the following month from 2.0–0.3 mg/kg per day and converted to an oral equivalent after return of bowel function. Mycophenolate is started immediately after surgery in order to reduce dependence on calcineurin inhibitors. Rapamycin can be added later. Anti-interleukin-2 receptor monoclonal antibodies, such as basiliximab, are sometimes used. Steroid-sparing regimens are becoming increasingly popular, but are not appropriate for patients with a history of autoimmune hepatitis.

COMPLICATIONS

Bleeding

Bleeding is more likely with a segmental graft or in the presence of graft dysfunction. Continuing hemorrhage despite correction of coagulopathy warrants early re-exploration, especially if there are signs of a developing abdominal compartment syndrome.

Primary non-function

This occurs in fewer than 5% of all transplants and requires emergency retransplantation. In such cases, signs of satisfactory early graft function (hemodynamic stability, early production of bile, normoglycemia, and resolution of lactic acidosis and coagulopathy) are absent.

Vascular thrombosis

Hepatic artery thrombosis (4–8% of cases) can be devastating. It is more common in smaller pediatric recipients. Various prophylactic agents, such as perioperative low-dose heparin and/or acetylsalicylic acid, are used and the packed cell volume should initially be maintained below 35%. Hepatic artery thrombosis may present in several ways: An acute deterioration in liver function progressing to fatal hepatic necrosis, an insidious onset with biliary complications or sepsis, or an absent arterial signal on routine Doppler US scan. If hepatic artery thrombosis is suspected within days of the transplant, urgent arterial computed tomodensitometry and/or laparotomy are warranted; if revascularization is not possible or unsuccessful, urgent retransplantation is usually required.

Portal vein thrombosis is uncommon (~2%). The narrow portal vein of children with biliary atresia is a risk factor. Early postoperative occlusion may present with acidosis and deranged clotting, gastrointestinal bleeding, ascites, or deteriorating liver function and demands urgent surgical correction. Late portal vein stenosis may cause portal hypertension (splenomegaly, variceal bleeding), but it can often be corrected by percutaneous transhepatic balloon angioplasty. In patients in whom percutaneous angioplasty fails to reopen the portal vein, it may be necessary to reconstruct the vein using a meso-Rex bypass vein graft. Less desirable, but often effective, a selective distal splenorenal shunt may be used to treat the complications of portal hypertension in late portal vein thrombosis.

Vena caval occlusion is very rare. Stenosis of the suprahepatic caval anastomosis leading to ascites and lower body edema is uncommon and can usually be treated by percutaneous angioplasty or stenting.

Biliary complications

The incidence of biliary leaks and strictures varies between 10% and 30% and is related to the type of graft, size of the duct, and number of bile ducts. Serious biliary complications are less common with whole organ grafts and more common in split-liver organs with more than one biliary anastomosis. Hepaticojejunostomies are more prone to leaks because of the relatively smaller size of the bile ducts and the nature of the anastomosis. Biliary complications are also related to arterial insufficiency.

Complications can be divided into early leaks and later strictures. Leaks from the cut surface of a split or a reduced-size liver may heal spontaneously, but a leak that persists for more than a few days should be repaired operatively and is likely to be from either the anastomosis or a segmental duct that was not seen at the time of the transplant. A bile leak that arises secondary to an arterial thrombosis is generally not repairable and requires retransplantation. Bile leaks present in a variety of ways: Bile in the drains after surgery, fever abdominal pain, or persistent jaundice. US or CT confirms a fluid collection near the cut surface posteriorly, or near the biliary anastomosis in the clinical setting suggestive of a bile leak.

Strictures generally appear after the first post-transplant month and can usually be dilated percutaneously by the interventional radiologists. Ischemic strictures may be isolated to the anastomosis or be widespread in the liver. Biliary strictures may be anastomotic or non-anastomotic (when they may reflect ischemia, preservation injury, chronic rejection, etc.). They present with jaundice and/or cholangitis. Strictures may present as Gram-negative sepsis in the absence of any other symptoms.

Endoscopic retrograde cholangiopancreatography (ERCP) with or without stenting can be a helpful adjunct in the management of small leaks or anastomotic strictures. Duct-to-duct anastomoses are more accessible, although specialized interventional gastroenterologists have now developed expertise in performing ERCP even in young patients with Roux-en-Y hepaticojejunostomies. Interventional radiologists and interventional gastroenterologists may also perform joined “rendez-vous” procedures to manage biliary strictures. Recurrent (more than two) anastomotic biliary strictures with resulting cholangitis requiring endoscopic or radiologic intervention should be revised surgically.

Acute cellular rejection

Acute cellular rejection typically manifests as an increase in the plasma levels of transaminases with or without a low-grade fever and irritability. An urgent Doppler US scan helps to exclude major surgical problems with the graft before performing a percutaneous needle liver biopsy to confirm the diagnosis. Acute rejection is initially treated with three daily doses of 10 mg/kg of IV methylprednisolone. Most patients respond within 48 hours. A further pulse of steroids may be given and/or other immunosuppressants introduced in resistant cases. Steroid-resistant rejection is treated with an anti-T cell antibody for 5–10 days, and an increase in the baseline immunosuppression.

Renal complications

Peritransplant renal dysfunction is common as a result of hepatorenal syndrome, ascites, cardiovascular instability, and nephrotoxic drugs. With good postoperative graft function, most cases recover well, although temporary renal support may be required.

Infections

Infectious complications are a major cause of mortality and morbidity after LT because of immunosuppression, indwelling catheters, pre-existing morbidity, malnutrition, and surgical complications. Early postoperative infections are mostly bacterial or fungal, but viral infections are particularly important later. Cytomegalovirus infection may cause fever, malaise, diarrhea, gastrointestinal bleeding, abnormal liver function, and pulmonary symptoms. Children are more likely to develop disease if they are primarily infected from a CMV-positive donor than when viral reactivation occurs in a previously immune recipient. Treatment with ganciclovir and reducing the immunosuppression are often effective. CMV prophylaxis is routinely given to naive recipients receiving a CMV-positive graft. Other opportunistic viral infections include herpes simplex, varicella zoster, adenovirus, and EBV. The last of these is a potential cause of post-transplant lymphoproliferative disease. Both CMV and EBV are to be monitored by serial estimations of viral load using the polymerase chain reaction.

Later complications, including rejection (especially from poor drug compliance), infections, side effects from immunosuppression, and graft dysfunction, continue to pose challenges. It is estimated that about 15% of children require retransplantation for complications at some stage.

OUTCOME

The United Network for Organ Sharing (UNOS) registry data for the United States show a 1-year overall patient survival of >90%, with graft survival exceeding 80%. Longer-term survival at 3 and 5 years is also in excess of 80–90%. Children with fulminant failure and encephalopathy or rapid decompensation of chronic liver disease have a worse prognosis primarily because of the comorbidities that accompany rapid decline in liver function. Pre-existing renal failure, vasopressor dependence, or stage 3–4 encephalopathy are also markers of worse outcome. A particularly problematic group are those children who weigh <5 kg at the time of LT with a diminished graft survival of about 80% at 1 year. Graft availability is often a problem and deterioration of other organ functions during the waiting period is a harbinger of a poorer prognosis.

With survival now exceeding 90% in most pediatric institutions, the emphasis has shifted to the quality of life enjoyed by the survivors. Large multi-institutional studies focus on the long-term impact of LT on school performances, psychosocial development and interactions, and quality of life indices.

FURTHER READING

Feier FH, da Fonseca EA, Seda-Neto J, Chapchap P. Biliary complications after pediatric liver transplantation: Risk factors, diagnosis and management. *World J Hepatol* 2015; 7: 2162–70.

Gurevich M, Guy-Viterbo V, Janssen M, et al. Living donor liver transplantation in children: Surgical and immunological results in 250 recipients at Universite Catholique de Louvain. *Ann Surg* 2015; 262: 1141–9.

Hong JC, Yersiz H, Busuttil RW. Where are we today in split liver transplantation? *Curr Opin Organ Transplant* 2011; 16: 269–73.

Meyers RL, Tiao G, de Ville de Goyet J, et al. Hepatoblastoma state of the art: Pre-treatment extent of disease, surgical resection guidelines and the role of liver transplantation. *Curr Opin Pediatr* 2014; 26: 29–36.

Sanada Y, Wakiya T, Hishikawa S, et al. Risk factors and treatments for hepatic arterial complications in pediatric living donor liver transplantation. *J Hepatobiliary Pancreat Sci* 2014; 21: 463–72.

89

Intestinal transplantation

JANG MOON AND KISHORE IYER

HISTORY

Intestinal transplantation is now an established treatment modality for intestinal failure. Following animal experiments in the 1960s, notably by Richard C. Lillehei at the University of Minnesota, David Grant in London, Ontario, who performed the first successful liver–intestinal transplant in the late 1980s. Since then, a small but active group of centers has reported increasing experience with the procedure. Deltz reported the first successful long-term small bowel transplant in an adult patient in 1988. Intestinal transplantation is now carried out by itself, i.e. isolated small intestine with or without colon, or as part of a composite graft with various organ combinations, combined liver and intestine, or a multivisceral graft with or without liver. Thus, conceptually it is useful to visualize the intestinal allograft within the context of multi-organ transplantation and to recognize that any allograft which includes the small intestine is defined and considered as an intestine transplant. In recent years, survival following intestinal transplantation has improved so that one-year patient survival at the major, experienced centers has outcomes similar to that of liver transplantation.

PRINCIPLES AND JUSTIFICATION

Parenteral nutrition is life-saving in intestinal failure due to short bowel syndrome and a variety of functional causes, such as dysmotility syndrome and pseudo-obstruction. The potential of a satisfactory and effective life-saving therapy in the form of parenteral nutrition distinguishes intestinal failure from other forms of end-organ failure. Uncomplicated parenteral nutrition in a stable patient population results in 5-year survival of close to 80%. A small subset of patients with refractory parenteral nutrition dependence are at risk of significant morbidity and even mortality due to complications that make it potentially dangerous or even impossible to continue this therapy. The most common of these complications is a syndrome of progressive cholestasis that may lead to end-stage liver disease. Despite satisfactory catheter-care techniques, some patients may also suffer inexplicably from central venous catheter-related complications in the form of recurrent life-threatening episodes of bacterial or fungal sepsis or loss of venous access due to venous thrombosis. The traditionally accepted indication for intestinal transplantation is the combined presence of refractory parenteral nutrition dependence and the onset of life-threatening complications of parenteral nutrition, described above. As clinical outcomes of intestinal transplantation improve, the indications have also been expanding. These include complete portomesenteric thrombosis in patients in need of liver transplantation, who cannot receive an isolated liver for technical reasons, non-resectable soft-tissue tumors at the root of the mesentery, extreme short bowel syndrome with gastrointestinal tract discontinuity making fluid–electrolyte management extremely challenging and even leading to poor quality of life in some cases. The current improved outcomes of intestinal transplant, not simply in terms of survival but also post-transplant quality of life, seem to justify these expanding indications.

PREOPERATIVE EVALUATION

Multidisciplinary evaluation is critical prior to a decision to proceed to intestinal transplantation. Key steps in this evaluation are:

- Evaluation of residual bowel anatomy
- Evaluation of residual bowel function/motility
- Demonstration of refractory parenteral nutrition dependence and inability to advance enteral feeds
- Evaluation of the presence and extent of liver disease
- Identification of other systemic illnesses/comorbid conditions
- Determination of any significant neurodevelopmental contraindications
- Determination of any psychosocial contraindications

It is our practice to carry out cross-sectional abdominal imaging, as well as appropriate gastrointestinal contrast radiographs if there appear to be opportunities for intestinal rehabilitation. Based on the clinical examination and biochemical tests of liver function, a liver biopsy may be required for accurate staging of the extent of liver disease. Recently, ultrasound elastography to measure liver stiffness has shown meaningful correlation with the results of liver biopsy, and, if validated, it provides a non-invasive method to monitor liver disease in this group of the patients. It is also important to evaluate the status of the central veins to ensure that central venous access can be securely maintained in the post-transplant period, at least in the short to medium term.

Such a detailed evaluation has allowed us to avoid the need for intestinal transplantation and even wean some patients off parenteral nutrition altogether. We have also on occasion uncovered previously undiscovered pathology, successful treatment of which has allowed patients to be successfully weaned off parenteral nutrition. On completion of evaluation, the appropriateness of intestinal transplantation for the patient can be determined with a high degree of confidence, and the choice of the type of graft (i.e. isolated small bowel, combined liver–small bowel, or multivisceral) can also be made in most cases. It is important to recognize that there is inevitably a long wait for a suitable deceased donor in most cases (particularly for recipients weighing <10 kg) and an accompanying high mortality rate on the waiting list, especially for patients awaiting combined liver–bowel transplantation. Therefore, early referral for children who appear to be failing parenteral nutrition may be an important step toward improving the outcomes for intestinal transplantation.

The choice of allograft is determined primarily by the presence or absence of advanced liver disease; the presence of cirrhosis with portal hypertension and massive hepatosplenomegaly mandates a combined liver–small bowel transplant. Lesser degrees of liver dysfunction may be reversible with good graft function from an isolated small bowel transplant. The indication for a multivisceral graft is more likely the necessity of total abdominal exenteration due to underlying abdominal pathology in addition to liver failure. The presence of irretrievably damaged foregut from enterocutaneous fistula or multiple prior procedures, or where there is no clear clinical benefit to preserve the native foregut would be a reasonable indication to proceed with multivisceral transplantation rather than combined liver–small bowel transplantation.

CHOICE OF DONOR

Hemodynamically stable donors, who are ABO-compatible and ideally about 30–80% of recipient weight, with evidence of good visceral perfusion (normal liver and renal function) and no acidosis, constitute satisfactory small bowel donors. Cytomegalovirus (CMV) mismatch between donor and recipient, particularly positive donor to negative recipient, appears to be a concern for the development of devastating CMV disease after transplantation. However, we no longer regard a CMV mismatch between donor and recipient (+ve to −ve) as a contraindication given current effective surveillance protocols and medical treatments to prevent or treat CMV disease.

Donor size, as a surrogate for allograft size, is a critical factor for successful intestinal transplantation. Most intestinal transplantation candidates have a history of repeated abdominal surgery and resulting loss of abdominal domain. This is often further compounded by loss of abdominal wall integrity. Adequate abdominal closure after intestinal transplantation is one of the most important technical factors for a successful outcome. For this reason, smaller donors are preferred in intestinal transplantation. There has been a long-standing practice to avoid donors who are less than 50% of recipient size. We have recently shown that, unlike in liver transplantation, "small for size syndrome" does not occur in isolated intestinal transplantation in the absence of liver allograft. As a general guide, we regard donor-to-recipient body weight ratios of 0.3 : 0.8 to be acceptable. For candidates who present with very limited abdominal domain and a difficult abdominal wall, donor-to-recipient body weight ratios as low as 0.3 : 0.4 would be suitable. Patients with functional disorders with few prior operations can accept larger donors. The initial enthusiasm for size-reduction techniques for both isolated bowel and combined liver–bowel grafting have been toned down by the continued challenges of sustaining good outcomes in this patient population, which can be adversely affected by any increase in technical risk.

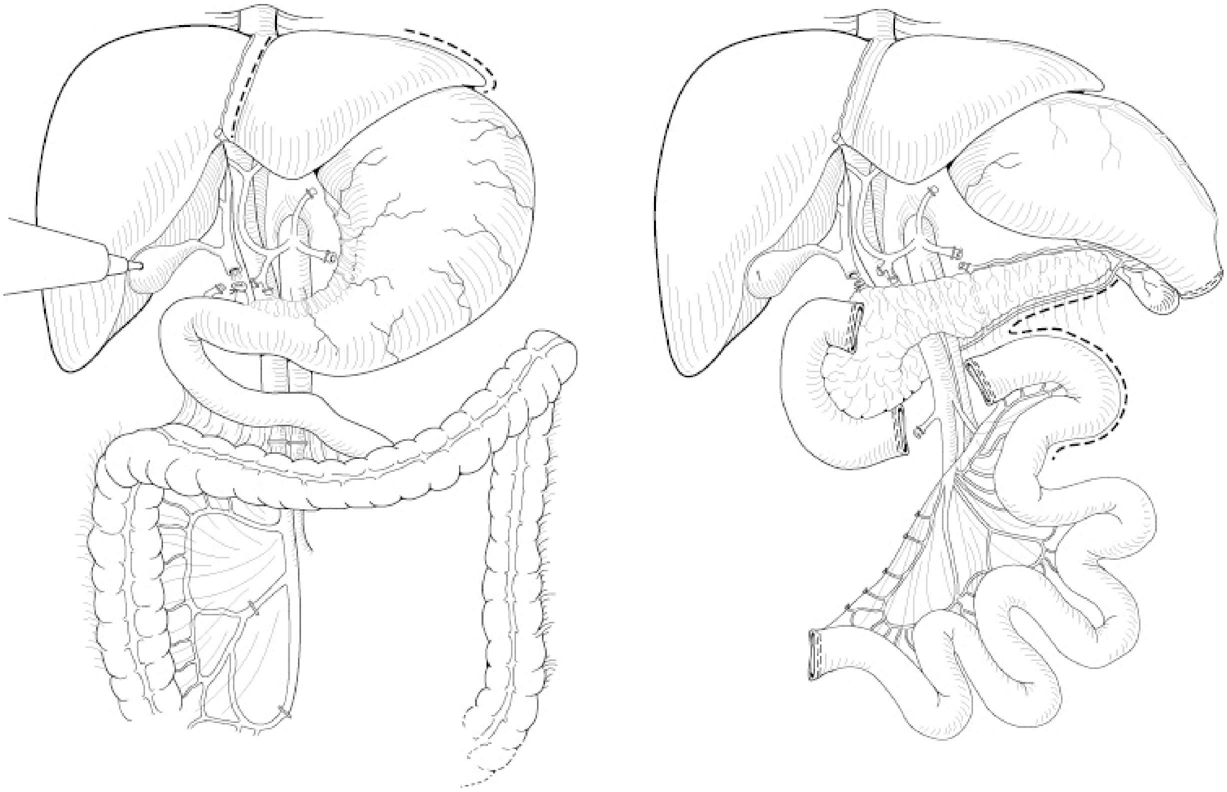
DONOR SURGERY

Routine preoperative antibiotics are administered to the donor. Other than maintaining good blood pressure and oxygen saturation, no other specific medical intervention is used during intestine recovery. Systemic heparinization is done with 300–500 IU/kg of IV heparin. Unlike some other groups, we no longer practice intestinal decontamination in the donor, with no apparent increase in complications. Large lymphoid tissue transfer from donor to recipient along with the intestinal allograft increases concern for development of graft versus host disease (GVHD) after intestinal transplantation. This may explain the apparent higher incidence of GVHD among pediatric recipients compared to other solid-organ recipients. Donor pre-treatment for lymphocyte depletion before cross-clamp has been proposed to prevent it. However, there is no clear scientific or clinical evidence to support this practice, and we no longer carry out donor re-treatment with depleting antibody.

The surgical technique for intestinal allograft recovery should be tailored to the choice of allograft organs. A flexible approach incorporating different options for necessary vascular structures and gastrointestinal tract allows optimal intestinal allograft recovery. One important feature of intestinal allograft recovery, regardless of type, is the need to recover various vascular grafts from the donor for possible vascular reconstruction on the backbench or recipient operative table. Of note, pediatric donors always have an inherent disproportion between upper and lower body. Because of this, unlike adult donors, iliac artery and vein from pediatric donors are usually not suitable as vascular conduits due to delayed development of lower extremities compare to the upper body. In this situation, size-compatible vascular grafts are the brachiocephalic trunk, and carotid and axillary arteries. Recovering thoracic aorta, superior vena cava, and subclavian vein are also important for possible vascular reconstruction.

Isolated intestinal allograft

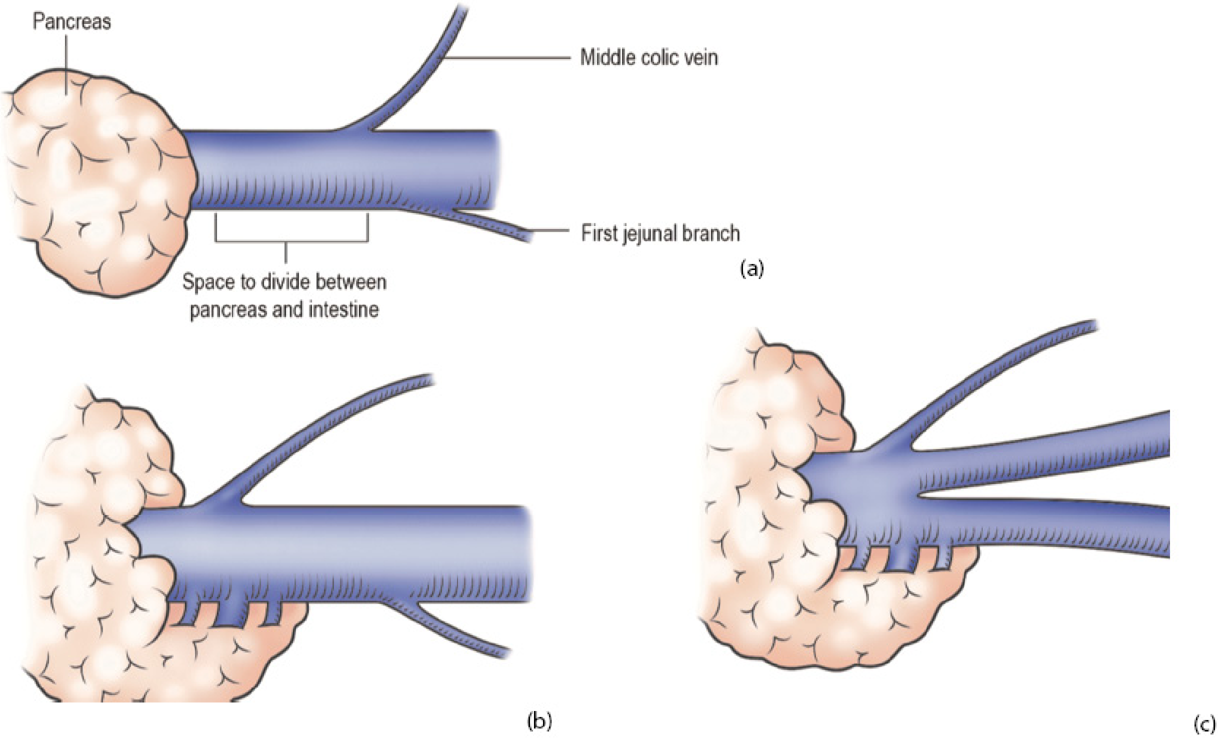
In our practice, the isolated intestinal allograft always includes a portion of proximal (right) colon in addition to entire small intestine (**Figure 89.1**). Arterial inflow is via the superior mesenteric artery (SMA) and venous outflow is through the superior mesenteric vein (SMV). Middle colic artery and vein are also included attached to the SMA and SMV respectively, when possible.



89.1

The procedure begins with a long midline incision and a median sternotomy. Right colon and small bowel mesentery are mobilized from the retroperitoneum until the left renal vein and the root of the SMA are identified. Omentectomy is performed from transverse colon to identify transverse mesocolon. The duodenum is separated from transverse mesocolon, which will expose the SMV at the inferior border of the third part.

This dissection will visualize the middle colic artery and vein along with the SMA and SMV. It is our routine practice to transplant proximal colon up to mid-transverse colon. To ensure abundant blood supply, middle colic artery and vein are included in the allograft whenever possible. Although it is thought best, it is not always possible to divide the pancreas and small intestine with preservation of all necessary vessels to both organs for subsequent transplant (**Figure 89.2a–c**). Small venous branches from the pancreatic head directly draining to the SMV are sometimes a problem in equitably dividing the vein between pancreas and intestine. Sometimes it is inevitable that these branches have to be sacrificed to obtain a small intestine allograft with adequate SMV drainage. Transverse mesocolon is divided distal to the middle colic artery. This dissection leads to the root of the SMA and SMV very easily. Proximal small intestine mesentery is a bit more difficult to divide due to many overlapping vascular arcades. For good hemostasis after reperfusion on the recipient, it is recommended to dissect these arcades in the donor before aortic cross-clamp. Attention should be given to preserve the first branch of the SMA and middle colic artery.



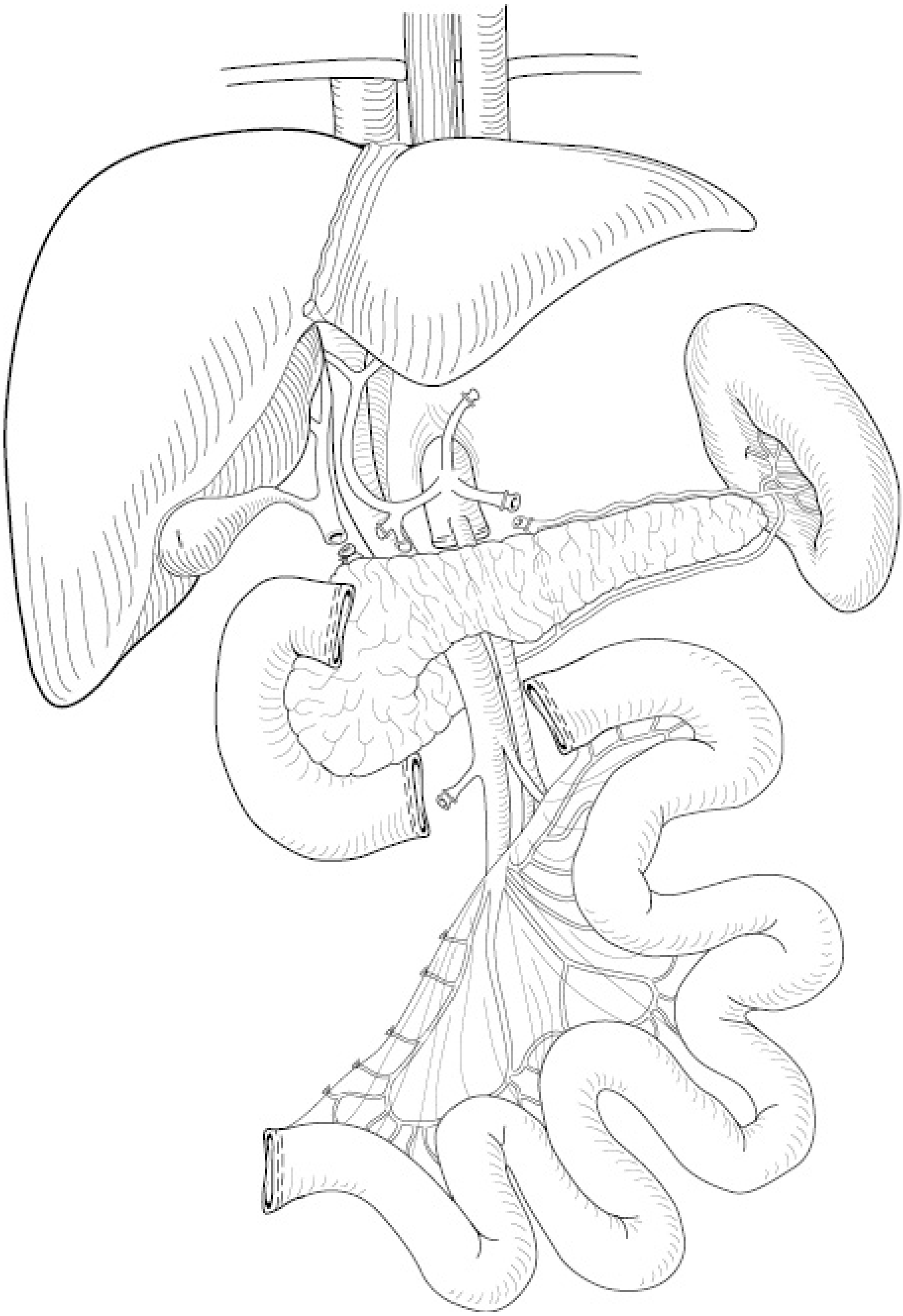
89.2a–c Example of pancreatic veins draining to the SMV. (a) Standard take-off of the middle colic vein and first jejunal branch from the SMV allows easy separation of pancreas and intestinal allografts. (b) High take-off of the middle colic vein – intestinal and pancreatic allografts can be separated but middle colic vein will be sacrificed. (c) High take-off of the middle colic vein and first jejunal branch – pancreas and intestinal allografts cannot be separated at procurement without critical compromise of venous drainage of one or both grafts.

Multivisceral allograft

Multivisceral allograft covers all the abdominal organs including stomach, pancreaticoduodenal complex, small and large intestine, and the liver. The allograft is recovered en bloc and treated as a single organ. Arterial inflow is via the abdominal aorta, which includes the celiac axis and SMA. Venous drainage is through the suprahepatic inferior vena cava (IVC), as in a liver transplant. The initial steps of the surgery are the same as for isolated intestinal allograft recovery through a mid-line incision with mobilization of right colon and small bowel mesentery. In addition, left colon mobilization is needed in multivisceral allograft recovery. The gastrocolic omentum is divided on the left approaching the splenic hilum. Splenic flexure of the colon is dissected and freed from the spleen. The splenorenal ligament is divided, mobilizing the spleen and tail of the pancreas toward the mesenteric root. The posterior aspect of the stomach is mobilized from retroperitoneal tissue with good hemostasis, extending this dissection to the esophagogastric junction. The distal esophagus is dissected and mobilized from adjacent structures, with good hemostasis to allow a satisfactory proximal gastrointestinal transection margin. The transverse mesocolon is divided as in isolated intestine allograft recovery. The hepatic hilum does not need to be exposed other than for biliary irrigation, through the gall bladder fundus before cold flush. Attention needs to be paid to the existence of the left replacement of the hepatic artery arising from the left gastric artery, which should be preserved carefully in the hepaticogastric ligament. The aorta is prepared for division just below the SMA root with preservation of the bilateral renal arteries on the renal allograft side.

Liver–small bowel allograft

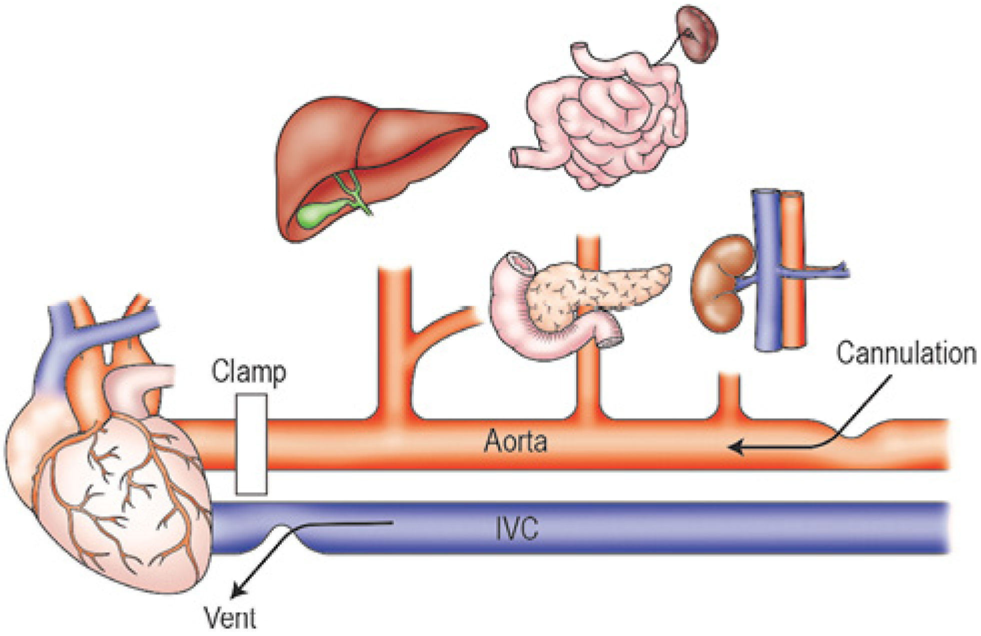
While the liver and intestine can be transplanted separately as individual organs, customarily they are transplanted en bloc. The pancreaticoduodenal complex can be preserved between the liver and intestine, with en bloc organ recovery, which is similar to recovering a multivisceral allograft, just without the stomach. Arterial inflow is through a segment of donor aorta bearing the celiac axis and SMA, exactly as in a multivisceral allograft. Usually, these arterial stumps are recovered in continuity with abdominal aorta. If recovered individually, the graft will require backbench vascular reconstruction to create a single arterial conduit using a “Y” graft. Venous outflow is via the suprahepatic IVC, as in a multivisceral graft. The spleen is removed on the back table. While, historically, the pancreas to the left of the hilar structures was transected and removed as shown in **Figure 89.3**, in combined liver–bowel transplant, we no longer remove the distal pancreas in any type of liver-inclusive allograft, whether combined liver–small bowel or multivisceral. Through a small opening in the fundus of the gall bladder, the biliary tree is flushed with saline before cold flush.



89.3

Cold flush

Intravenous heparin (300–500 IU/kg) is administered by the anesthesiologist. A cannula is placed in the abdominal aorta immediately above the bifurcation (**Figure 89.4**). Portal flush via the inferior mesenteric vein does not seem to confer any benefit and we no longer use an inferior mesenteric/portal flush. The thoracic organ recovery team sections the IVC above the diaphragm to vent the blood and clamps the aorta. The supraceliac or thoracic aorta is clamped simultaneously wherever it is feasible. Cold preservation solution (e.g. University of Wisconsin solution) is administered via the aortic cannula. The amount of cold preservation solution flush depends on the size of the donor, and is approximately 50 mL/kg. Cold slush is distributed in the abdominal cavity to achieve rapid core cooling of the organs.



89.4 The concept of cold flush of abdominal viscera and procurement of organs singly or as a "cluster."

Back-table preparation

The purpose of backbench surgery is not only to prepare vascular stumps for future anastomosis and to ensure hemostasis but also to reconstruct artery and vein for safe and efficient recipient surgery. Size matching vascular grafts is essential. Identifying all potential bleeding points on the back table is critical, particularly at the mesenteric root. Control of bleeding from the root of the mesentery after allograft reperfusion in the recipient can be very difficult and dangerous. Appendectomy, cholecystectomy, and, if the denervated stomach is included, pyloroplasty, can be done on the back table based on the surgeon's preference.

Preparation of isolated intestinal allograft

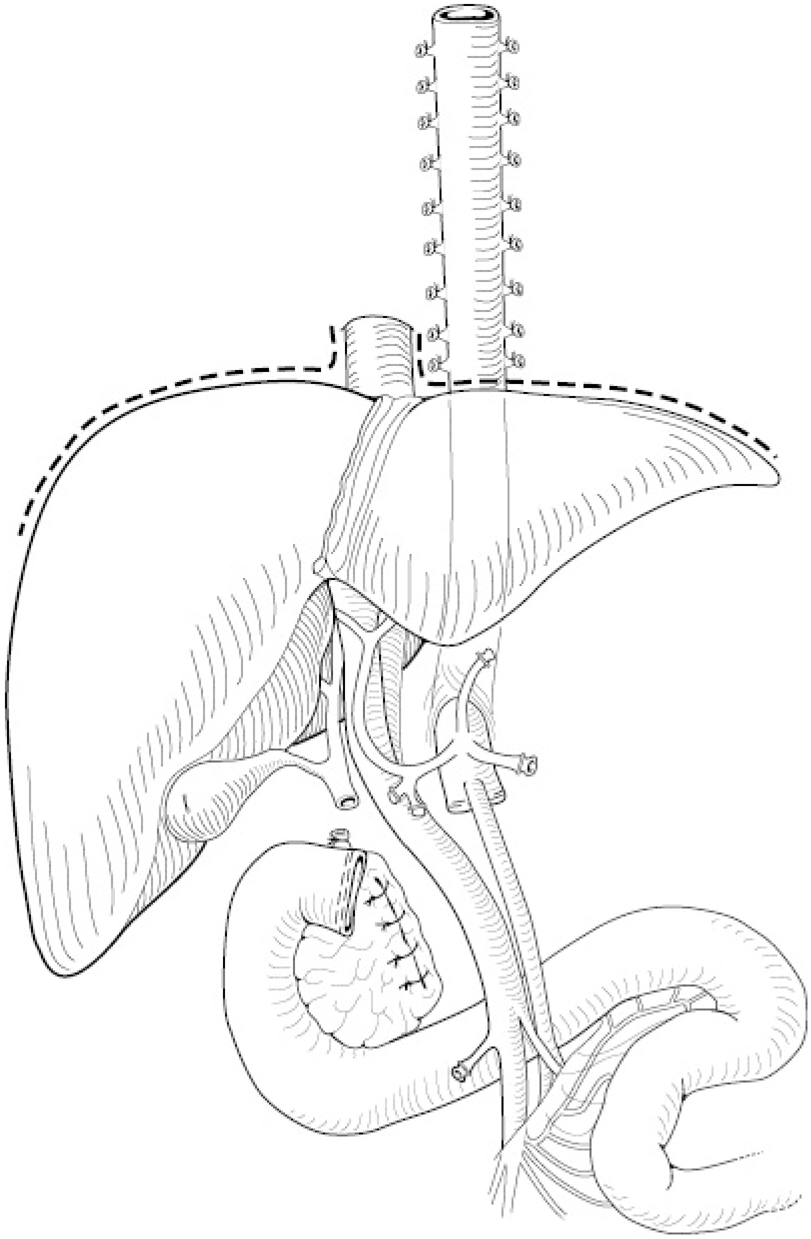
The SMA is dissected free from the periadventitial tissue, progressing from its aortic take-off. Attention should be given to avoid injury to SMV branches, crossing the SMA. If the pancreas is not recovered for the purpose of transplant into a different recipient, the stump of the inferior pancreaticoduodenal artery needs to be ligated. Further dissection is not needed below the inferior pancreaticoduodenal artery. The middle colic artery (if recovered) and the first jejunal branch should be preserved. The SMV stump should be carefully oriented to avoid any torsion in the recipient surgery. The splenic vein stump can be used as an anatomical marker at the left edge. If middle colic vein is recovered, in the anatomical position, it should be on the anterior surface of the SMV. It is our practice to mark right and left edges of the SMV with Prolene™ stitches. If any vascular stump is too short for safe anastomosis, we recommend placing extension vascular conduits with size-compatible vascular grafts on the back table to facilitate easy and safe anastomosis.

Preparation of the liver–small bowel or multivisceral allograft

In most cases, thoracic and abdominal aorta is recovered with the celiac axis and SMA *in situ*. The posterior surface of the allograft is exposed in the ice-cold preservation solution basin. Four corner stitches are placed at each end of the aorta. Surrounding soft tissue is sharply dissected and the aorta is mobilized with preservation of all lumbar branches. All lumbar branches are ligated with individual silk ties. When dissection approaches the root of the celiac axis and SMA, extreme care needs to be taken to avoid surgical injury to each artery.

The posterior surface of the liver is prepared in the same manner as for liver allograft back-table preparation. The preparation of the liver starts with clearing the diaphragmatic tissue and skeletonizing the IVC. The right adrenal vein should be identified and ligated. Diaphragmatic and pericardial tissue is sharply removed, with identification of each phrenic vein. All branches should be ligated with individual silk ties. If there is left replacement hepatic artery from the left gastric artery, it should be carefully preserved in the hepaticogastric ligament.

We recommend placing an extension aortic graft on the distal abdominal aorta. This is divided just below the SMA to protect both renal arteries during donor surgery. A segment of donor thoracic aorta can be used as an extension graft (**Figure 89.5**). Special attention should be paid not to alter or narrow the ostium of the SMA. Placing an extension graft at the inferior end allows the recipient surgeon the flexibility to use either side of the aorta for arterial inflow.



89.5

RECIPIENT OPERATION

Completion of enterectomy: Reclaiming domain and preparation for vascular anastomoses

Abdominal incision can be vertical or horizontal, depending on prior operative scars. A bilateral subcostal incision, with suitable midline extensions (cruciate if necessary) is preferable if a liver allograft is included, and is often essential for a multivisceral transplant. A configurable and versatile self-retaining retractor, such as the Thompson™ abdominal retractor, facilitates exposure. Extensive adhesiolysis is often required. Prior bowel anastomosis between the remnant small bowel and colon is taken down. Obviously non-functional or dilated remnant bowel is excised. If transplant includes liver allograft, entire viscera can be removed en bloc, including liver. Right and left colon mobilization is performed. The mesenteric root is exposed and ready to clamp. Hepatic hilum dissection is not required in isolated intestinal transplantation.

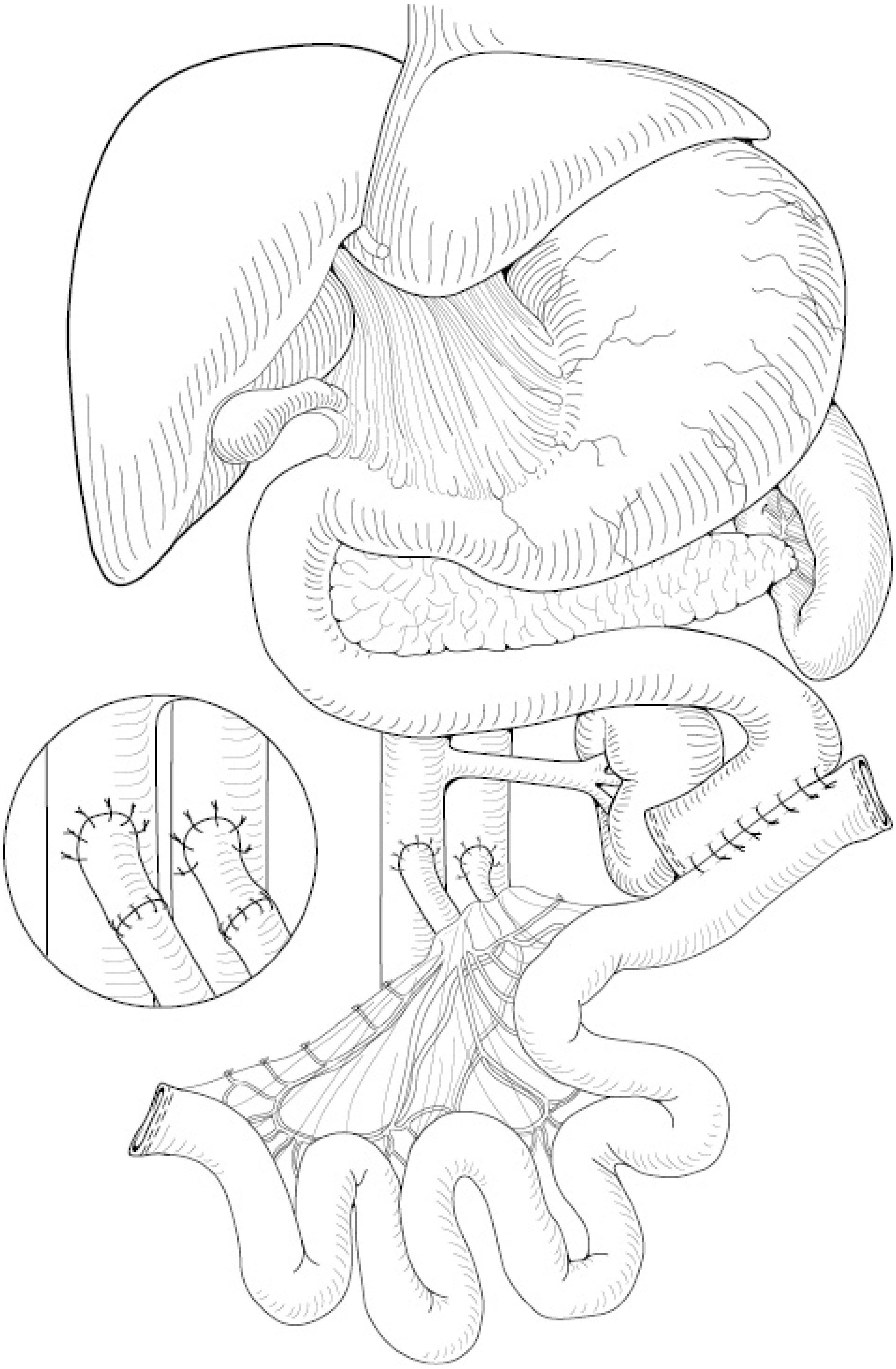
For liver-containing grafts (liver–small bowel or multivisceral), the liver is mobilized from diaphragmatic tissue. Both cava resection (with subsequent replacement) and preservation (for piggyback organ implantation) can be used at the surgeon's discretion. Hepatic hilar dissection and hepatectomy are similar to the corresponding steps in isolated liver transplantation (See Chapter 88). It is important to note that, for multivisceral transplantation, especially in the presence of confluent portomesenteric venous thrombosis where the operation is often performed, it is preferable to treat the entire abdominal viscera as a single organ to be excised en bloc, rather than to attempt a very bloody and dangerous hilar dissection.

It is critically important to preserve arterial inflow and venous drainage for the remaining native viscera. Except in the event of total eviscerectomy for multivisceral transplantation, the root of the SMA should be preserved in addition to the celiac axis. When the recipient liver is removed and native foregut is left *in situ,* we perform a portocaval shunt between the end of the native portal vein and the infrahepatic IVC. This allows for decompression of all remnant native foregut viscera, such as the stomach, duodenum, and pancreas. The IVC is then reclamped above the shunt. Where prior abdominal exenteration removes native foregut in the abdomen, including a near-total gastrectomy prior to a multivisceral transplant, the need for a portocaval shunt is obviated. Vasculature of distal colon needs close attention, particularly with respect to the inferior mesenteric vein. The inferior mesenteric artery is easy to identify and preserve. However, the inferior mesenteric vein can be easily injured during completion enterectomy, which will result in significant venous congestion of the left colon. If the inferior mesenteric vein is not preserved, we recommend trimming down native colon to the rectum, with venous drainage to the hemorrhoidal complex.

Bilateral ureters are easily involved and potentially injured in surgical dissection and special attention should be paid to protect the ureters. In selected cases, preoperative ureteral stent placement may help avoid accidental ureteric injury.

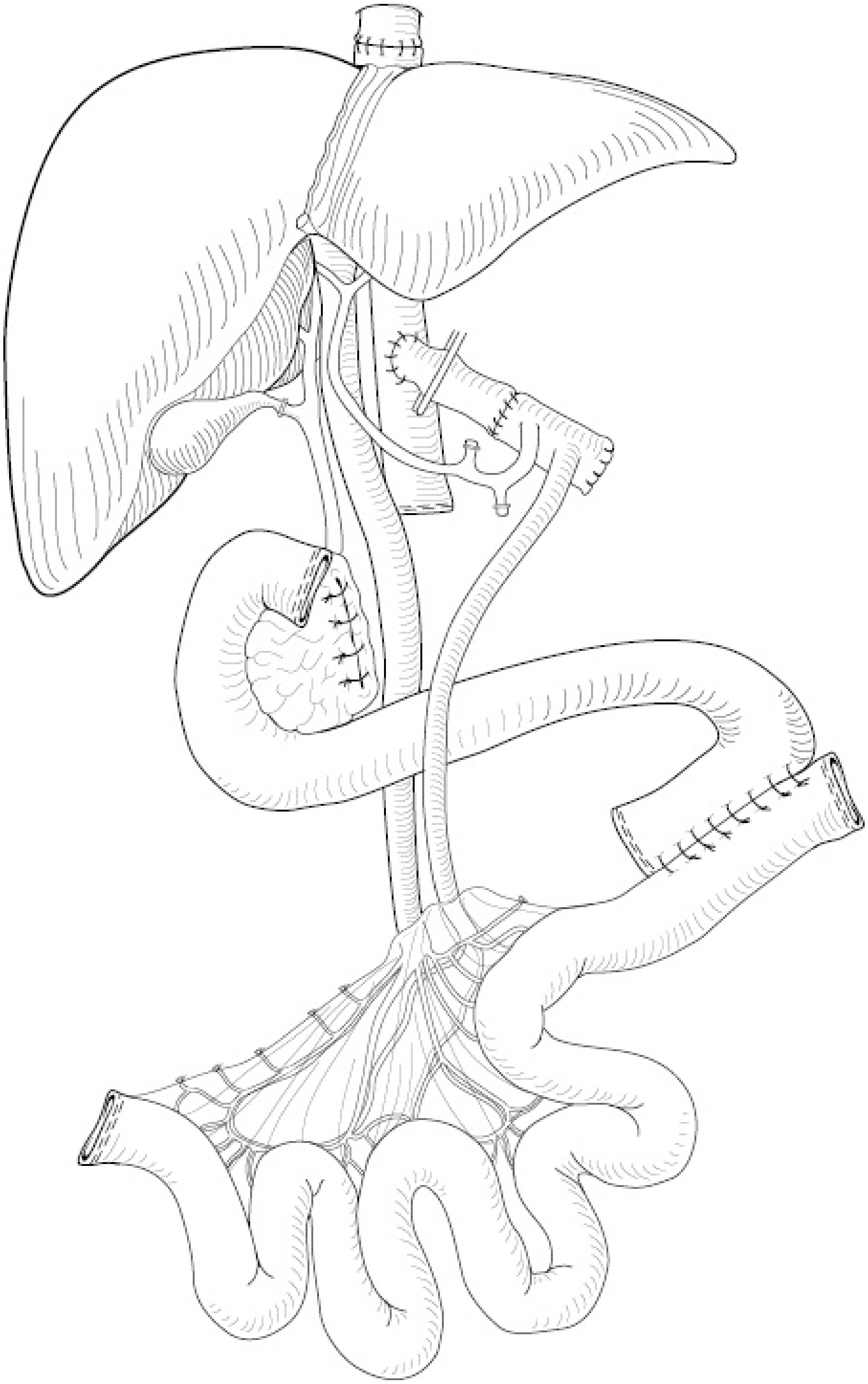
Implantation of the allograft: Vascular and gastrointestinal anastomoses

We prefer to use the infrarenal aorta and IVC of the recipient to vascularize the graft. Extension vascular conduits on both structures make graft vascular anastomosis easy and safe without increased vascular complication. Vascular clamps are placed in side-biting fashion when possible, but in the small recipient these are invariably occlusive. Size-compatible artery and vein are chosen and standard end-to-side vascular anastomosis is performed to recipient infrarenal aorta and IVC (**Figure 89.6**). End-to-end vascular anastomosis is performed between extension vascular conduit and corresponding allograft vascular stumps. The SMA of the graft is anastomosed initially, via the arterial extension to the infrarenal aorta of the recipient, followed by anastomosis between the SMV of the graft and the IVC, via an extension graft.



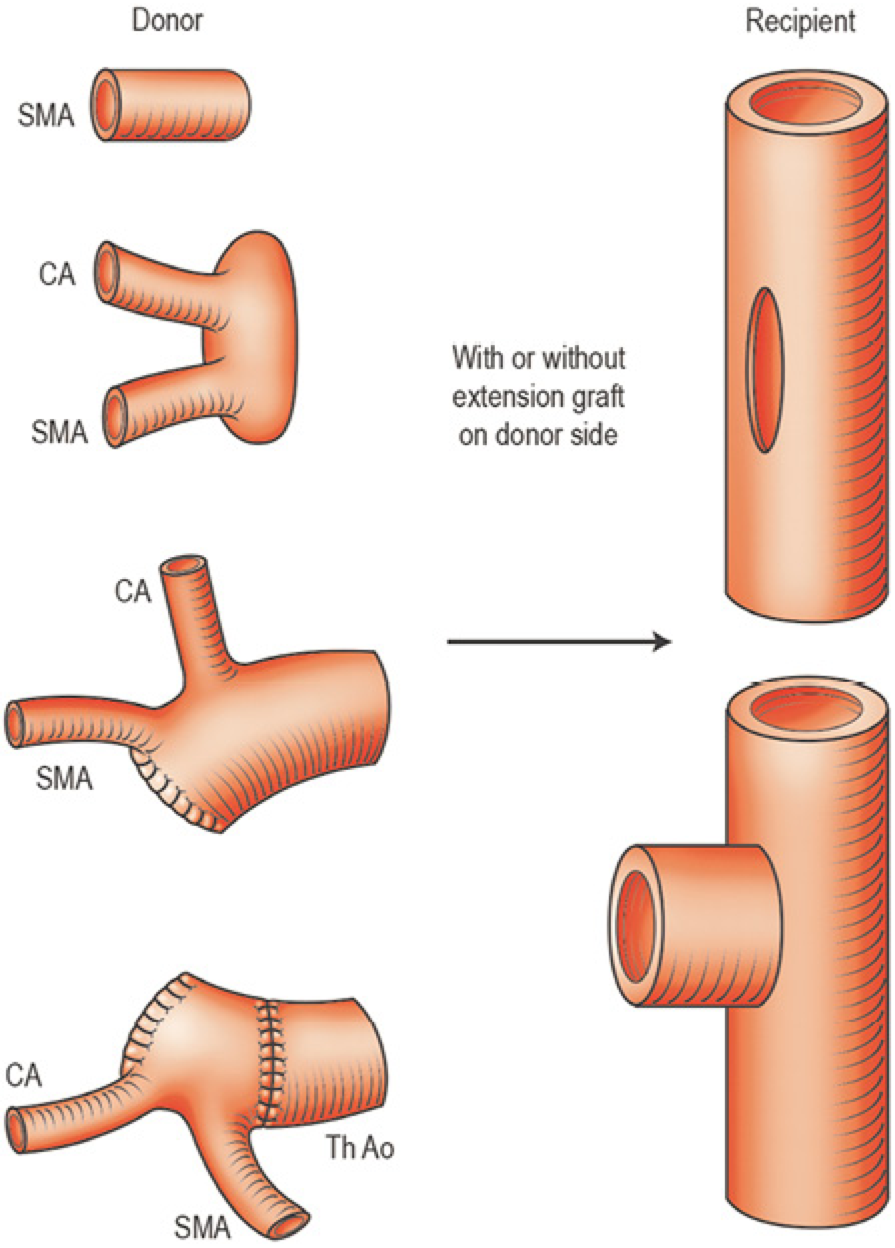
89.6

Due care must be taken to ensure that the venous anastomosis is cephalad to the arterial anastomosis, and that proper orientation is maintained (**Figure 89.7**). In multivisceral and liver–small bowel transplantation, venous anastomosis is between the suprahepatic IVC of the allograft and the recipient vena cava below the right atrium, as in isolated liver transplantation. Systemic venous drainage of the allograft (donor SMV to recipient IVC) is not physiologic since venous gut drainage would bypass hepatic circulation. However, this technique is actually relatively easy and is without serious complications after transplantation. Thus far, it has had no detectable impact on patient or graft survival. The use of an interposition graft in this manner also allows a convenient aortic anastomosis in the depths of the wound, without having the composite allograft in the way, and allowing final arterial anastomosis to the liver–small bowel or multivisceral graft at the level of the wound (**Figure 89.7**).



89.7

Reperfusion starts with release of the venous clamps. Bleeding points in the cut edge of the mesentery are controlled, followed by release of the arterial clamps. Once hemostasis is secured, attention is turned to restoring intestinal continuity. We carry out side-to-side (functional end-to-end) anastomosis between the proximal end of the graft and the remnant native proximal jejunum or duodenum (**Figure 89.8**). Similarly, distal anastomosis is carried out between the graft colon stump and the distal native remnant bowel. In the case of a multivisceral transplant, anastomosis is carried out between partial donor stomach and a very small remnant of native stomach at the gastroesophageal junction or to the native esophagus itself. We perform anastomoses with a standard two-layered technique due to the theoretically higher risks of poor healing in an immunosuppressed patient.


Donor
Recipient
SMA
CA
SMA
With or without extension graft on donor side
CA
SMA
CA
Th Ao
SMA

89.8 Potential donor and recipient vascular conduits for arterial inflow to the allograft. (SMA, superior mesenteric artery; CA, celiac axis; Th Ao, thoracic aorta.)

Completion of transplant surgery: Stoma creation and abdominal wall closure

In the absence of reliable biomarkers to monitor allograft health, it is conventional practice to create a stoma for the purpose of frequent endoscopy and biopsy surveillance of the intestinal allograft. Distal ileum is used to create the stoma. A loop ileostomy, reverse Bishop–Koop (Santulli), or double-barrel stoma is brought out.

Abdominal wall closure can be challenging from abdominal wall scarring due to previous multiple abdominal surgeries and small recipient abdominal domain with a relatively large allograft. It is critically important to achieve at least skin cover of the intestinal allograft. If primary abdominal wall closure is not possible, extensive abdominal wall plasty, devascularized deceased donor fascia, or even abdominal wall transplant may be indicated. Of note, any synthetic mesh should be avoided due to extremely high risk of infection and eventual risk for enteric fistulization. We have resorted to skin closure alone in difficult cases, with definitive repair of an often large incisional hernia at a much later date.

Gastrostomy or jejunostomy tube insertion can be carried out surgically if indicated.

POSTOPERATIVE CARE

Nasogastric decompression is maintained for 5–7 days after transplant, introducing clear liquids with return of peristaltic activity. Changes in color and appearance of the stoma or its output are a cause for concern and reflect graft vascularity early after transplant. Perioperative anti-infectives are maintained for at least 1 week after transplant. Various prophylactic agents against fungi, viruses such as CMV, and opportunist infectious agents such as *Pneumocystis jiroveci* (previously *P. carinii*) are routinely prescribed for periods of between 3 months and 1 year after transplant.

Most centers use an induction regimen with antibody followed by maintenance immunosuppression with tacrolimus and steroids in the long term. Close monitoring of tacrolimus levels is essential for the first few months after transplant to maintain optimal levels.

With satisfactory stoma activity, fat-free diets or enteral formulas are introduced about 7–10 days after transplant and advanced gradually based on tolerance and stoma outputs. Transition to complete formulas and/or liberalization of fat in the diet is delayed until 4–6 weeks postoperatively to allow time for lymphatic reconstitution after transplant. Oral aversion is a common obstacle for transitioning to standard diets in the longer term. The ability to remove the central line after showing full enteral tolerance is a major milestone in these patients. A very small percentage of patients continue to require supplemental IV fluids until stoma takedown between 6 months and 1 year after transplant.

In the absence of meaningful biochemical markers for rejection, endoscopy and biopsy of the allograft still remain the gold standard to diagnose rejection. Sudden increment of stoma output, changes of stool characteristics (watery or bloody), fever of unknown origin, or bacteremia are important clinical features suggesting rejection. Diagnosis of acute rejection should be made strictly on the basis of histology in addition to supportive clinical features. Most episodes of acute rejection occur in the first year after intestinal transplantation. Treatment with larger doses of methylprednisolone is the first line of treatment for biopsy-proven rejection. Severe or steroid-resistant rejection needs antibody treatment. Graft removal with the ability to discontinue immune suppression and return to parenteral nutrition should be considered to avoid mortality if rejection persists.

COMPLICATIONS AND OUTCOME

Despite the magnitude of the operative procedures described above, the postoperative management remains the most challenging aspect of the care of these patients. The relatively high incidence of acute rejection, now about 30–40% in a lymphoid-rich graft, requires high levels of immunosuppression compared to other solid-organ transplants. This in turn leads to a high incidence of bacterial, viral, and fungal infections that require close monitoring and aggressive early treatment. Bacterial infections are often due to enteric Gram-negative organisms, which may have to be treated while maintaining significant immunosuppression. In the longer term, complications related to immunosuppression, such as post-transplant lymphoproliferative disease related to Epstein–Barr virus, and chronic rejection continue to pose serious challenges to patient and graft survival.

Despite these challenges, the results of intestinal transplantation continue to improve, allowing patient survival at 1 year of the order of 90–95% and graft survival of 80–85%. With improving outcomes, survival at 5 years is now >65%. The vast majority of survivors enjoy enteral autonomy off parenteral nutrition. When one recognizes that these outcomes are achieved in patients who have “failed” parenteral nutrition and whose survival without transplant approaches zero, it is hoped that earlier patient referral before the onset of advanced liver disease and better selection may allow continued refinement and even better outcomes.

RECENT DEVELOPMENTS

Our center, along with a few others, now routinely includes the right half of the colon as part of the allograft in patients who come to transplant without a colonic remnant. Without an appreciable increase in complication rate, initial impressions suggest a favorable impact on fluid and electrolyte management and preservation of renal function.

Loss of domain in the patient with a short bowel with a complex surgical history remains a major technical challenge. Autologous tissue reconstruction, while desirable, is often not possible, and synthetic mesh repairs are accompanied by unacceptable infection rates. Experience with composite abdominal wall transplants remains limited, but there is some favorable experience with the use of avascular donor abdominal fascia; the key to success with the use of fascia appears to be meticulous removal of all fat and muscle and prevention of desiccation of the fascia during the back-table preparation. Our group has had a consistent and high level of success, employing meticulous skin closure alone in cases of limited domain with a large graft, and elective repair of the hernia a year or two later under more controlled conditions.

Recent success with the use of IV fish oil emulsions and other lipid-sparing strategies, and indeed improved understanding of parenteral nutrition-associated liver disease, has brought in its wake new challenges for intestinal transplantation. Reversal of biochemical cholestasis in the patient with extreme short bowel, who may still have advanced histological liver injury, creates a therapeutic dilemma in terms of choice of organs for the patient being considered for intestinal transplantation. Current allocation systems do not offer any prioritization for histological liver disease, forcing an often difficult choice between a prolonged wait for a combined liver–small bowel graft or an isolated intestinal transplant with unknown but presumably higher risks of liver decompensation.

We have over many years been impressed with the general philosophy that, to the extent possible, less may be better in intestinal transplantation, reflecting the complicated surgical histories, infection risks, and burden of immunosuppression in these patients. This has led us to cautiously move away from certain established principles in this challenging field. The following are examples of our own paradigm shifts in management:

- Routine acceptance of donor-to-recipient weight ratio <0.5, and often as low as 0.3.
- Routine acceptance of CMV-positive donors for CMV-negative recipients.
- Shunning a practice of routine protocol surveillance endoscopy and biopsy, in favor of our current protocol of endoscopy and biopsy only "for cause." Our preliminary data suggest that the addition of routine surveillance protocol endoscopy and biopsy in the absence of signs or symptoms of graft dysfunction confers no survival advantage to patients or grafts.
- Given the move away from surveillance endoscopy, avoidance of stomas in most cases.

While more data need to be accumulated to confirm the validity of these changes in practice, our current patient survival of 95–100% at 1 year and commensurately high graft survival, with a length of hospital stay that is currently an average of 21–28 days, down from historic stays of 45–60 days, suggests that isolated intestinal transplantation for intestinal failure could indeed soon be considered first-line therapy for patients with intestinal failure, even in the absence of parenteral nutrition-related complications.

FURTHER READING

Fishbein T. Intestinal transplantation. *N Engl J Med* 2009; 361: 998–1008.

Moon JI, Schiano TD, Burnham A, et al. Small-for-size syndrome does not occur in intestinal transplantation without liver containing grafts. *Transplantation* 2018; 102(8): 1300–6.

Moon JI, Schiano TD, Iyer KR. Routine surveillance endoscopy and biopsy after isolated intestinal transplantation – revisiting the gold standard. *Clin Transplant* 2019; 33(10): e13684.

Starzl TE, Todo S, Tzakis A, et al. The many faces of multi-visceral transplantation. *Surg Gynecol Obstet* 1991; 172: 335–44.

Sudan DL. Treatment of intestinal failure: Intestinal transplantation. *Nat Clin Pract Gastroenterol Hepatol* 2007; 4: 503–10.

Todo S, Tzakis AG, Abu-Elmagd K, et al. Intestinal transplantation in composite visceral grafts or alone. *Ann Surg* 1992; 216: 223–34.

90

Interventional radiology

DEREK J. ROEBUCK

HISTORY

Since its origins in the 1980s, pediatric interventional radiology (IR) has expanded to become an essential part of hospital pediatrics. Most pediatric IR has been adapted from adult practice in an attempt to improve on existing surgical techniques, and radiologists now perform many procedures that were previously exclusively in the domain of pediatric surgery.

PRINCIPLES AND JUSTIFICATION

Most pediatric IR procedures are based on very simple concepts. In one, a structure is first accessed by percutaneous puncture with ultrasound (US) and/or fluoroscopic guidance. A guidewire is inserted, the needle is removed, and a catheter is introduced into the structure over the guidewire (the "modified Seldinger" technique). The procedure is named after Swedish radiologist Sven-Ivar Seldinger (1921–1998). Invented in 1952, it can be summarized in his words: "Needle in, wire in, needle off, catheter on wire, catheter in, catheter advance, wire off." This concept is the basis of angiography, abscess drainage, biliary drainage, nephrostomy, cecostomy, and one method of gastrostomy. Another concept is dilation, which is performed by crossing a stenosis with a guidewire followed by a balloon catheter, and inflating the balloon, usually with the use of fluoroscopic guidance. This is the basis of angioplasty (of arteries and veins) and dilation of tracheobronchial, ureteral, and esophageal and other gastrointestinal strictures.

In all cases, the intention is either to replace surgical operations with less invasive and potentially more effective and/or less costly treatments, or to provide treatments for which there is no surgical alternative.

PREOPERATIVE CARE

In general, the risk of bleeding from IR procedures is extremely small, even in the presence of moderate coagulopathy or thrombocytopenia. On the other hand, percutaneous techniques offer less opportunity for intraoperative recognition and treatment of bleeding complications than the corresponding open, endoscopic, or laparoscopic procedure. It is therefore good practice to check coagulation values and platelet levels before most procedures in the lungs, pleural space, and abdomen, where postoperative bleeding is more likely to be a significant problem.

Uncorrectable coagulopathy is a relative contraindication to some procedures (e.g. liver or renal biopsy) but not others (e.g. peripherally inserted central venous catheters). Blood is usually cross-matched only for procedures where sudden, potentially catastrophic bleeding might occur (e.g. angioplasty).

SEDATION AND ANESTHESIA

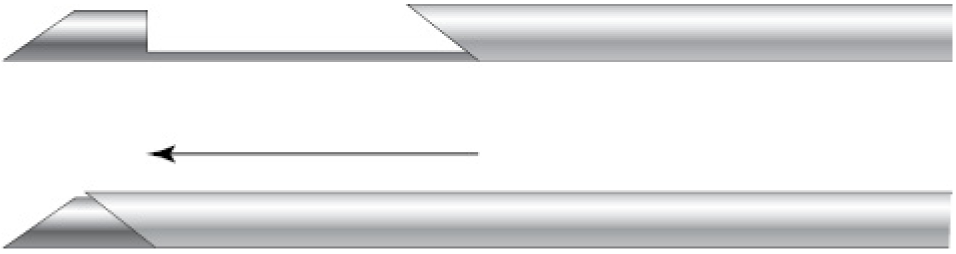
Although a wide range of anesthetic practice still exists in pediatric IR, there is a clear trend away from radiologist-managed IV sedation toward general anesthesia (GA), which is less unpleasant for the child and may make the procedure safer. GA is, of course, essential for procedures that are very painful or very long, or require the patient to remain very still (e.g. digital subtraction angiography).

When GA is not used, topical anesthetic cream is applied before the procedure to the area where local anesthetic will be injected. If the procedure is performed under GA, a long- acting local anesthetic is injected to optimize postoperative pain relief.

For brief but painful procedures, such as renal biopsy or abdominal abscess drainage, older children can breathe an equimolar mixture of oxygen and nitrous oxide (Entonox™). This is very effective when used in combination with topical and local anesthetic.

BIOPSY AND DRAINAGE PROCEDURES

Image guidance makes percutaneous biopsy and drainage safer and quicker, and blind needle biopsy is now appropriate only for the occasional superficial lesion. In children, nearly all abdominal biopsies can and should be performed with US guidance rather than computed tomography (CT), because US has better spatial and temporal resolution (and is therefore safer) and is much quicker. Percutaneous biopsies in children are usually performed with semi-automatic core needles (**Figure 90.1**). The cutting tip is advanced to expose a slot in which tissue is trapped by an outer sleeve, which advances when the needle is fired. These devices are much easier to use than manual biopsy needles (e.g. Trucut™) and biopsy gun systems. Fine-needle aspiration with immediate cytological examination may be appropriate for some indications (e.g. thyroid nodules).



90.1

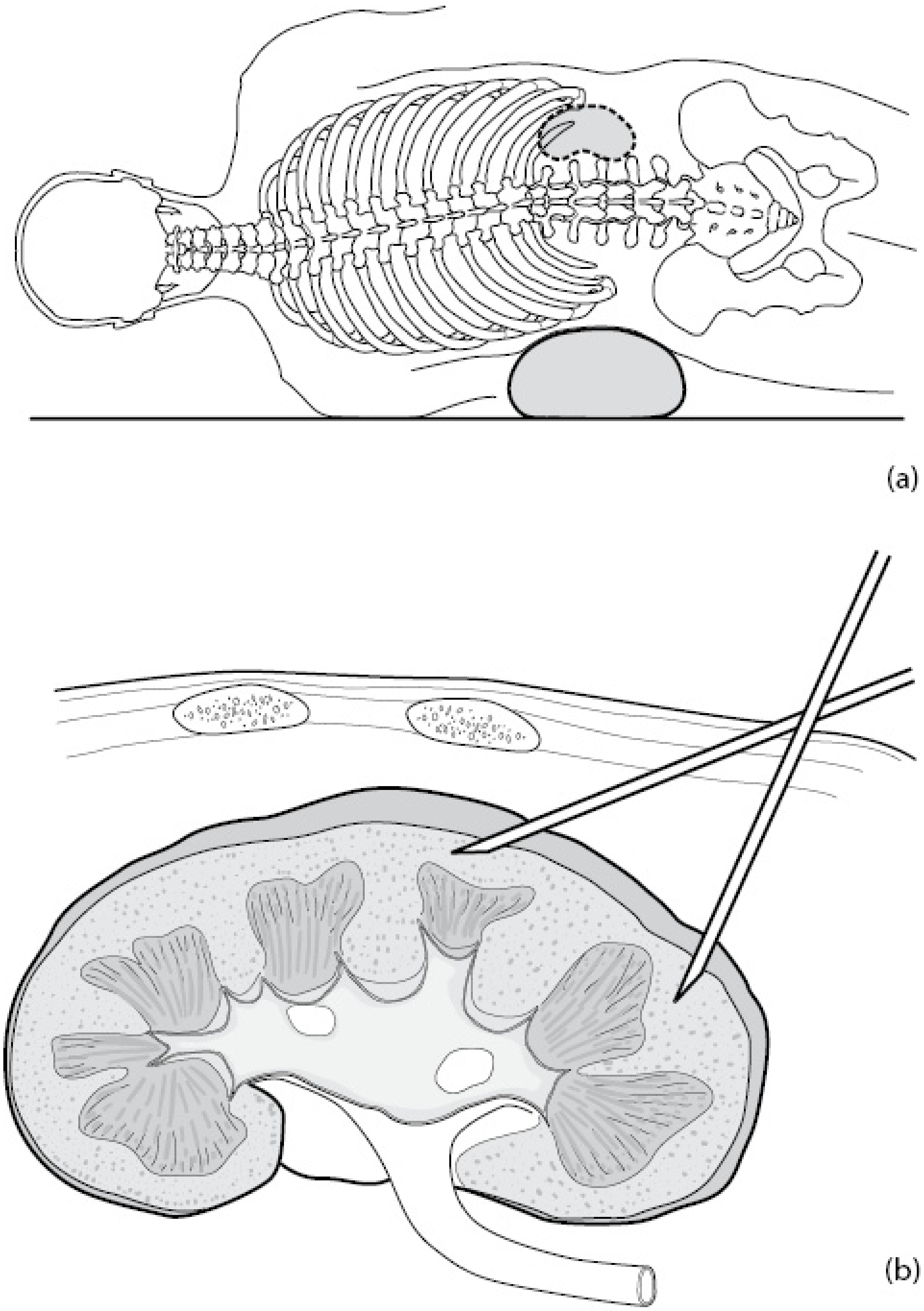
Renal biopsy

INDICATIONS

The most common indications for renal biopsy are nephrotic syndrome and impaired function following renal transplantation.

POSITIONING AND ANESTHESIA

Renal biopsy is performed under GA in young or uncooperative children, and with sedation or Entonox™ in older children. Either kidney can be biopsied. The procedure can be performed with the patient prone or seated, but it is easier to place the patient in a decubitus position. This makes it much easier to administer Entonox™, and, when GA is used, avoids the need for endotracheal intubation and muscle relaxants. A large pad is placed under the patient's abdomen to increase the convexity of the side to be biopsied (**Figure 90.2a**). Renal transplant biopsy is performed in the supine position, and does not usually require GA or sedation because it is much less painful than biopsy of a native kidney.



90.2a,b

TECHNIQUE

The tract is infiltrated with local anesthetic down to the level of the renal capsule. A 16-gauge semi-automatic core biopsy needle is then inserted through a short stab incision (**Figure 90.2b**). If the child is able to cooperate, they are asked to stop breathing as the needle is advanced under real-time US guidance into the kidney and the biopsy is taken. A shallow or deep trajectory may be appropriate, depending on the size of the kidney and the relationship of its lower pole to the ribs. When the child is under GA, suspension of breathing is not required, and the anesthetist may prefer to allow the patient to breathe spontaneously. A pathology technician is present to examine the core and confirm that the sample contains sufficient glomeruli. If not, another core is taken. Using this method, a definitive histopathology report should be possible for 99% of biopsies.

COMPLICATIONS

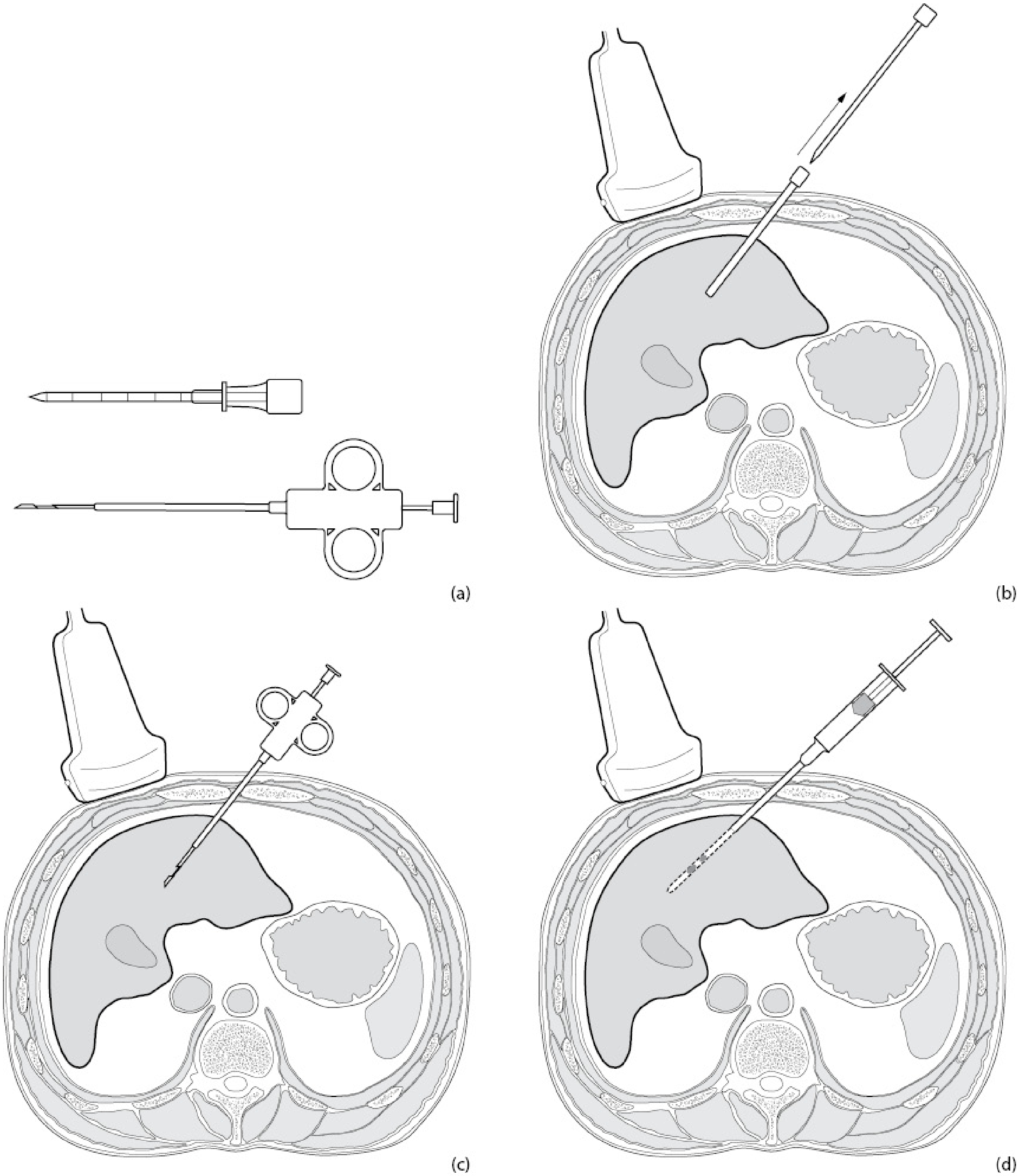
Significant complications, such as hematuria requiring transfusion, or injury to other organs, should occur in less than 2% of biopsies. The most important complication is arteriovenous fistula. These lesions tend to resolve spontaneously but require embolization (see **Figure 90.10**) if they have clinical consequences such as hematuria or hypertension.

Liver biopsy

Percutaneous liver biopsy can be used to evaluate generalized liver disease, large liver masses, or small focal lesions. In all these cases, we prefer to use US guidance.

TECHNIQUE

A coaxial technique is used for almost all liver biopsies. For generalized liver disease, we use a 17-gauge outer needle, which accepts an 18-gauge biopsy needle (**Figure 90.3a**). A subcostal approach is almost always possible, and this avoids the risks of pneumothorax and hemothorax associated with the traditional intercostal approach. The outer needle is advanced into the liver under US guidance, taking care to avoid the gall bladder and major vessels (**Figure 90.3b**). Its trocar is then removed and replaced with the semi-automated core biopsy needle (**Figure 90.3c**). After obtaining three or four cores of tissue (**Figure 90.3d**), the tract is occluded, usually with gelatin foam plugs, in order to prevent intraperitoneal bleeding.



90.3a–d

COMPLICATIONS

The most important complication is bleeding, which can occasionally be life-threatening. Frequent meticulous postoperative observations are essential, and should be continued for several hours. Hemorrhage requiring treatment (by blood transfusion, transarterial embolization, or emergency surgery) is rare. It is more common in children with uncorrected coagulopathy or thrombocytopenia, and those who have undergone bone marrow transplantation. Transjugular biopsy has been recommended in these patients to try to reduce the risk of post-biopsy bleeding, although it has not been proven to be safer than percutaneous biopsy.

Tumor biopsy

INDICATIONS

The histologic diagnosis of most tumors outside the central nervous system is made by US-guided needle biopsy. Our experience is that needle biopsy is probably safer than, and equally accurate to, open surgical biopsy. The period of convalescence is shorter, and this may allow earlier commencement of chemotherapy. Laparoscopic biopsy is a useful alternative for certain thoracic and abdominal tumors. Open biopsy is performed when it is considered to be safer, or when other techniques have failed.

TECHNIQUE

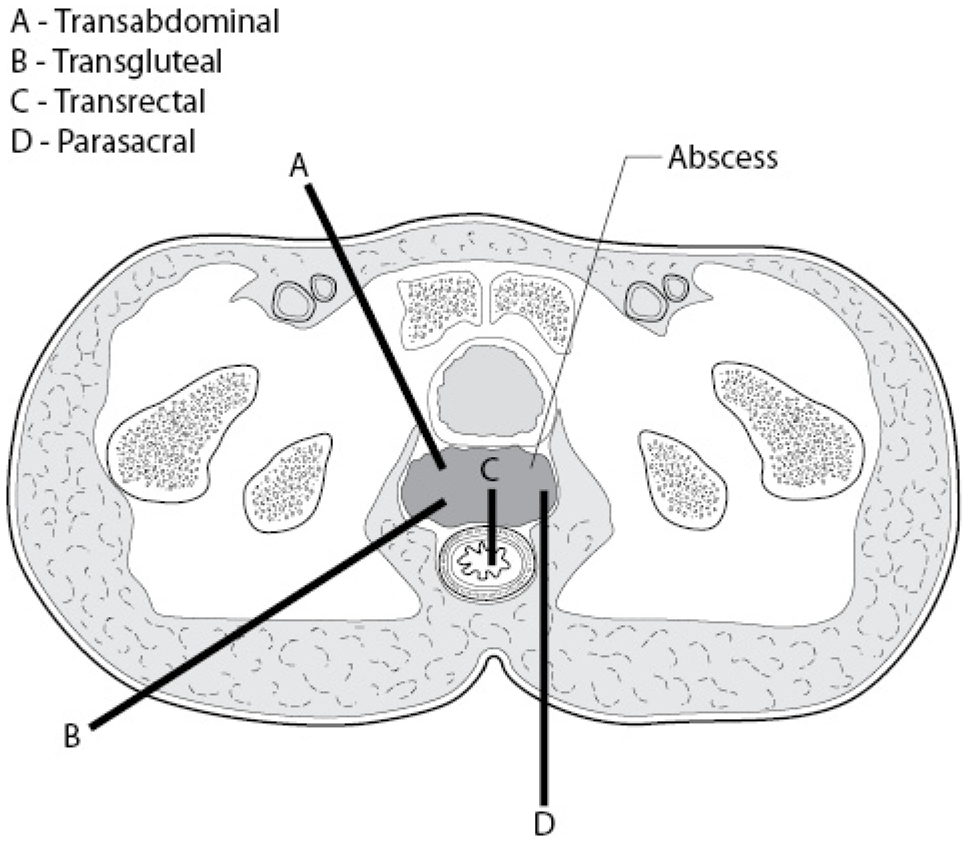
We use CT mainly for very small retroperitoneal lesions, especially in older children, and lung lesions that do not reach the visceral pleura. Recent advances in pediatric pathology, including immunohistochemical and molecular techniques, have improved the accuracy of needle biopsy. In most patients, US-guided tumor biopsy can be performed at the same GA as insertion of a central venous access device and, where appropriate, bone marrow aspirates and trephines. The coaxial technique (see “Liver biopsy”) is particularly useful for tumor biopsy, because numerous cores of tissue can be obtained from different parts of the mass, using a single puncture of the tumor capsule. Plugging the tumor biopsy tract may prevent seeding of tumor cells.

COMPLICATIONS

Complications are very rare. Inadequate biopsies can be avoided by careful preoperative imaging, and it is usually worth delaying the biopsy for a day or two to obtain all relevant imaging before the procedure.

Aspiration and drainage of collections

Most fluid collections can be drained by IR techniques. In the upper abdomen, most collections are approached through the anterior abdominal wall. Certain lesions, such as urinomas or pancreatic collections, are drained retroperitoneally. Pelvic collections can be approached through the anterior abdominal wall or the rectum using US guidance (**Figure 90.4**).



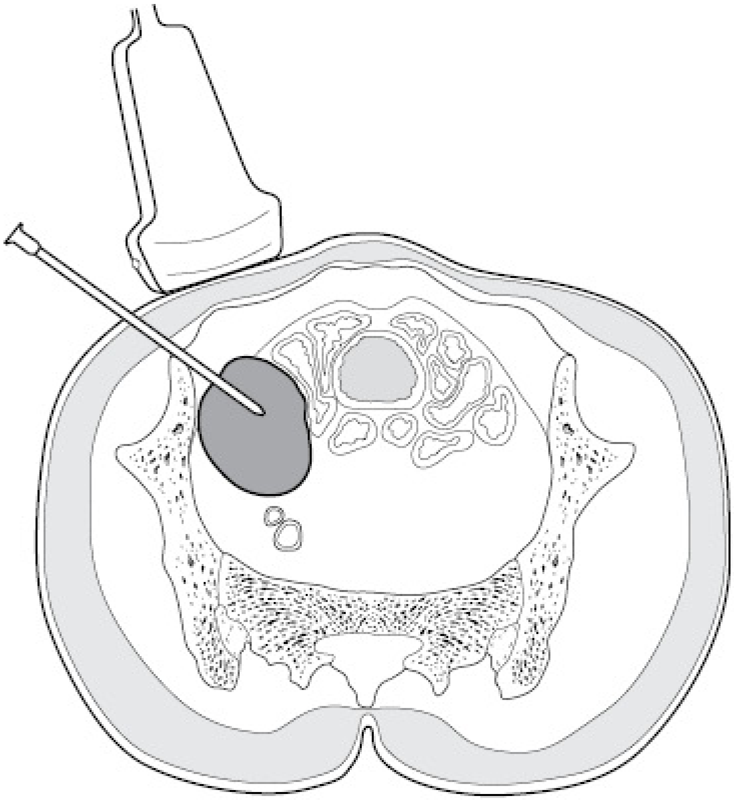
90.4

TECHNIQUE

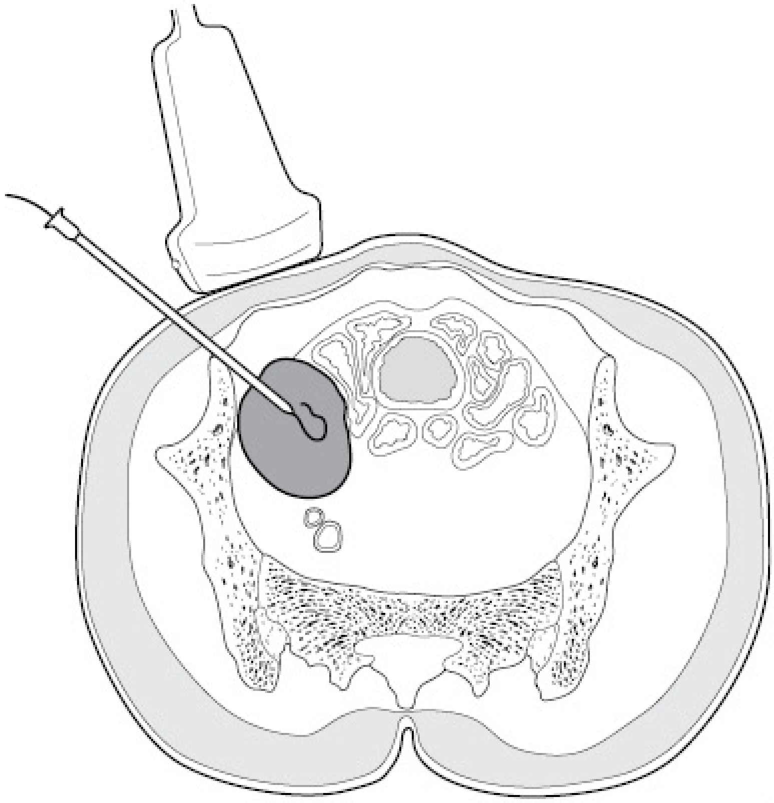
The usual technique is based on US-guided puncture of the fluid collection. Guidewire insertion and positioning of the drainage catheter can be monitored with US or fluoroscopy if necessary. The first step is to select a safe path to the collection by careful US examination. After injection of local anesthetic, a short stab incision is made in a skin crease, and the collection is punctured using real-time US guidance. The author uses a trocar needle (usually 18-gauge) for this purpose, because the trocar can be withdrawn slightly into the blunt outer needle, which can then be advanced atraumatically past bowel loops if necessary. When the tip of the needle is in the collection, the trocar is removed and fluid aspirated and saved for culture (**Figure 90.5a**). A guidewire of appropriate diameter is inserted (**Table 90.1**) (**Figure 90.5b**). The best guidewire has a stiff shaft, with a floppy segment about 30–60 mm long at its tip. The tip coils in the collection first, and then increased resistance is felt, indicating that the stiff part of the guidewire is in the collection. The needle can then be removed and the tract dilated over the guidewire (**Figure 90.5c**).

Table 90.1 **Guidewires and their diameters**

Needle gauge	Guidewire diameter
21	0.018 inch (0.46 mm)
20	0.025 inch (0.64 mm)
19	0.035 inch (0.89 mm)
18	0.038 inch (0.97 mm)



(a)



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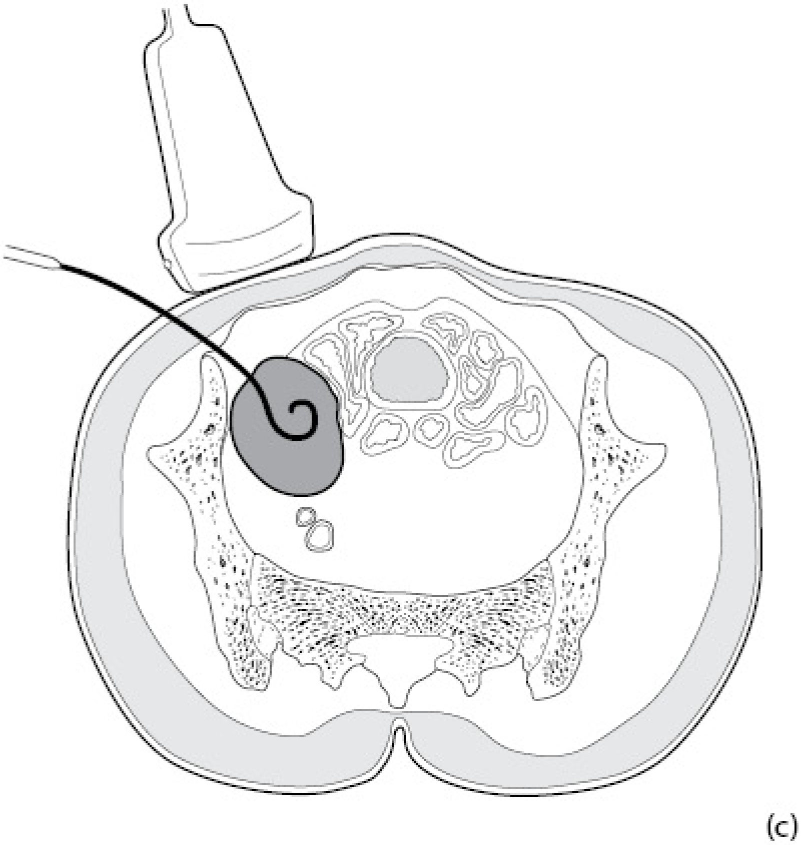


90.5a–c

The author often uses a dilator 0.5–1 Fr larger than the intended drainage catheter at this stage. The pigtail catheter (with its internal stiffening cannula) is then advanced over the guidewire and into the collection. The guidewire and stiffener are withdrawn and the pigtail is formed and locked. (There are different methods for this, according to the design of the catheter.) Fluid is aspirated to confirm adequate position, and a drainage bag is connected. The catheter can be sutured to the skin or retained with a highly adhesive dressing.

For transrectal drainage, the needle can be laid along the operator's index finger, with the inner trocar partly withdrawn. When the finger is inserted into the patient's rectum, it is easily seen with transabdominal US, especially if the bladder is full, enabling puncture of a pelvic collection with real-time guidance.

GASTROINTESTINAL INTERVENTION

Esophageal dilation

Fluoroscopically guided balloon dilation of esophageal strictures is simple and safe, and has replaced bougienage. Endoscopy can be regarded as an additional procedure, with certain advantages over a purely fluoroscopic technique, including the ability to inspect and biopsy the esophagus and to evaluate the stomach and proximal small intestine.

INDICATIONS

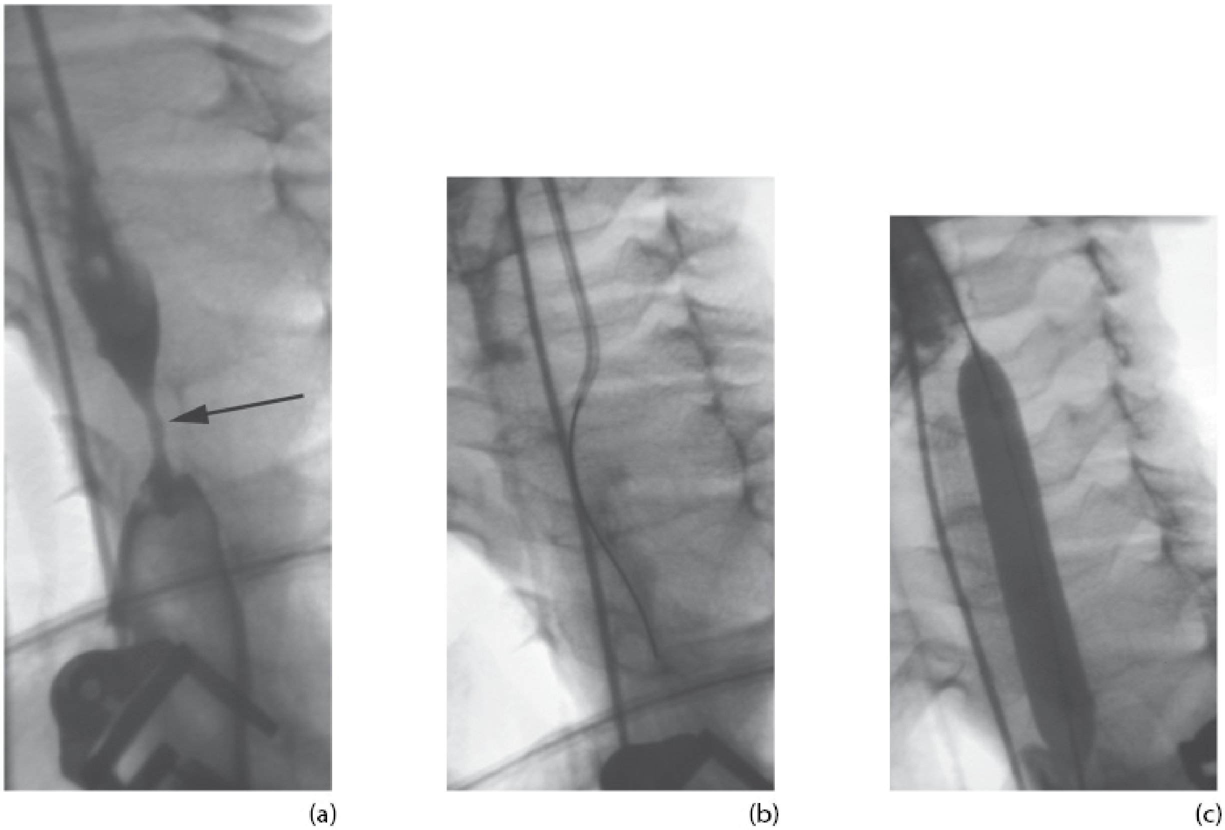
Common causes of esophageal stenosis in children include congenital and surgical strictures, ingestion of corrosive substances, gastroesophageal reflux, achalasia, and dystrophic epidermolysis bullosa. The natural history, response to treatment, and probability of complications all depend strongly on the underlying cause of the stricture. Repeated dilation is often necessary, especially in children with corrosive injury and epidermolysis bullosa. Resection of esophageal strictures (except for congenital stenosis) is now rarely necessary.

ANESTHESIA

Although it is possible to dilate the esophagus in a sedated child, we prefer to use GA, because it is less distressing for the child and minimizes the risk of a severe vagal reaction. Endotracheal intubation avoids the severe airway compression that may otherwise occur during balloon dilation of esophageal strictures in small children.

TECHNIQUE

Most strictures can be treated with balloon dilation using an antegrade (transoral) approach. A catheter with a gently curved tip is inserted into the upper esophagus, using fluoroscopic guidance if necessary, and a contrast study is performed (**Figure 90.6a**). The stricture is then crossed with a guidewire, which is advanced to the stomach. The catheter is removed, and replaced with an angioplasty catheter with a balloon of appropriate diameter. The angioplasty catheter is advanced over the guidewire until the radio-opaque markers on the balloon straddle the stricture (**Figure 90.6b**). The balloon is then inflated with dilute radiographic contrast (not air) (**Figure 90.6c**). A pressure inflation device is not required, but fluoroscopy is essential to confirm that the stricture has been successfully dilated, as shown by abolition of the waist on the balloon. The balloon is then deflated, and the catheter and guidewire removed. Occasionally, it is impossible to cross the stricture from above. When this occurs in children who have a gastrostomy, a retrograde (transgastric) approach is almost always successful. In children whose strictures recur relentlessly after dilation, mitomycin application and/or temporary stenting can be tried.



90.6a–c

COMPLICATIONS

Complications occur most often during dilation of corrosive strictures, followed by those associated with gastroesophageal reflux. Three types of esophageal injury occur. Guidewire perforation of the esophagus can be recognized at fluoroscopy and treated conservatively. Submucosal tears of the esophagus are seen as contained extravasation of contrast alongside the inflated balloon. These also do well with conservative treatment. Full-thickness perforation during inflation of the balloon is uncommon but requires treatment with analgesia, nasojejunal or parenteral feeding, and broad-spectrum antibiotic therapy. This is usually successful, but surgical repair is sometimes necessary.

Other gastrointestinal dilation

Balloon dilation may also be used to treat strictures elsewhere in the gastrointestinal tract including the pylorus, duodenum, colon, and rectum.

Gastrostomy

Radiologic gastrostomy can be performed either by an antegrade technique, in which the G-tube is pulled down the esophagus into the stomach, or a retrograde technique, in which it is pushed into the stomach through the anterior abdominal wall. The antegrade technique is essentially the same as percutaneous endoscopic gastrostomy, except with fluoroscopic guidance. The retrograde technique may be useful in children with a narrow or diseased esophagus. The perioperative care is the same as for endoscopic gastrostomy.

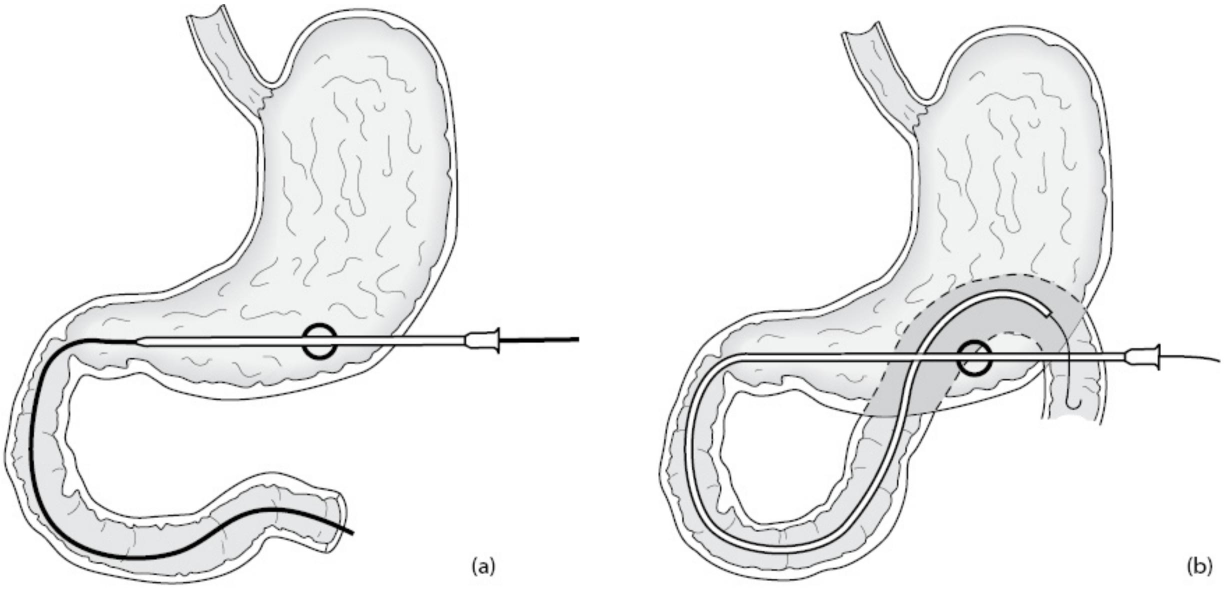
TECHNIQUE

The antegrade and retrograde techniques are similar. We usually give the child a small amount (2–3 mL/kg) of barium sulfate suspension orally or by nasogastric tube the evening before the procedure, to opacify the colon. US is used to mark the position of the left lobe of the liver. A puncture site is selected, where possible lateral to rectus abdominis, about 2 cm from the left costal margin. Occasionally, anatomic considerations (e.g. scoliosis) require the use of a different puncture site. Local anesthetic is infiltrated and a short transverse incision is made in the skin. For antegrade gastrostomy, a snare is passed into the stomach through the mouth. The stomach is then inflated with air. This can be done via the snare catheter or a separate nasogastric tube. In children with esophageal atresia, the stomach can be inflated after percutaneous US-guided puncture with a 25-gauge needle. When the stomach is inflated, an 18-gauge trocar needle is inserted through the incision and used to puncture the stomach. This must be done quickly, before the air escapes from the stomach into the small bowel. The author finds biplane fluoroscopy useful at this point, but it is not essential. Some operators use IV glucagon (40 µg/kg, maximum 1 mg) to delay gastric emptying. The snare in the stomach is opened, and a 0.035-inch (0.89 mm) guidewire is advanced through the needle and grasped with the snare. The wire is then carefully pulled out of the mouth, and the tract dilated to permit the retrograde passage of the snare catheter through the abdominal wall to the mouth. The snare catheter is then used to pull the lubricated gastrostomy tube down the esophagus, and out through the anterior abdominal wall. The procedure is otherwise the same as for endoscopic gastrostomy (see Chapter 6).

For retrograde gastrostomy, the stomach is punctured in the same manner as for the antegrade technique, but a needle preloaded with a suture anchor device may be used instead of the trocar needle. These temporary retention devices are used to hold the stomach against the anterior abdominal wall as the tract is dilated. A locking pigtail catheter or balloon device can then be inserted. Although these are more easily dislodged than flanged gastrostomy catheters, they are easier to remove or exchange when the tract is mature. If necessary, a transgastric jejunal feeding catheter may be inserted at the time of creation of the gastrostomy.

Transgastric jejunal intubation

This requires the passage of a transpyloric tube through a pre-existing gastrostomy, to a position distal to the duodenojejunal flexure, using fluoroscopic guidance. A stiff dilator (9–15 Fr) is used to engage the pylorus, and a floppy-tipped hydrophilic guidewire is advanced into the duodenum (**Figure 90.7a**). The dilator is removed, and a long catheter is inserted over the guidewire and manipulated across the duodenojejunal flexure (**Figure 90.7b**). The floppy-tipped wire may then be exchanged for a stiff wire, if necessary, and the catheter removed. The transgastric jejunal tube is then lubricated (internally and externally) and inserted over the stiff wire. Injection of contrast through the catheter will confirm its position. Coaxial systems, in which a transpyloric tube can be advanced through a gastrostomy catheter, are also available.



90.7a,b

UROLOGIC INTERVENTION

Most urologic interventions are based on nephrostomy. In children, this is usually straightforward when performed with US guidance, except when the pelvicalyceal system is not dilated. The nephrostomy tract can also be used for other interventions, such as percutaneous stone removal or antegrade ureteral dilation and stenting.

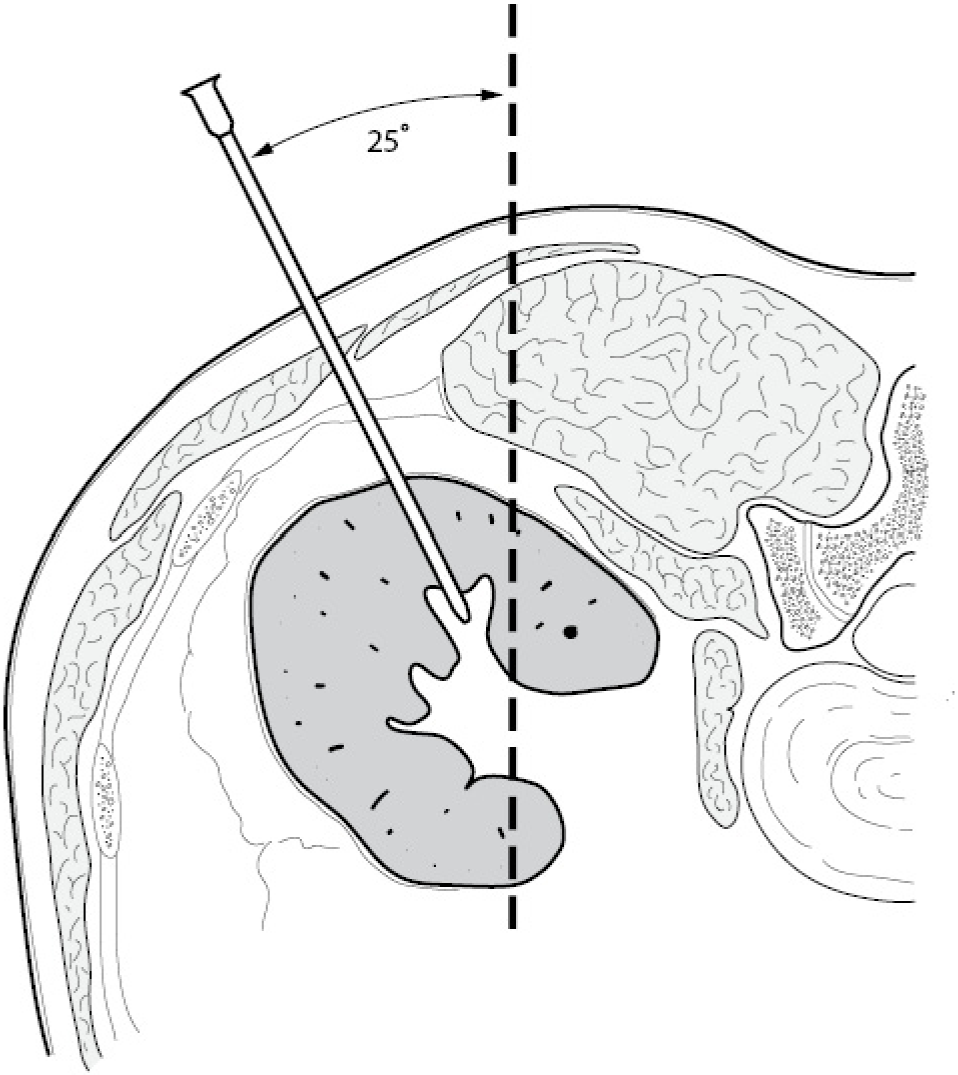
Percutaneous nephrostomy

INDICATIONS

The most common indications for nephrostomy in children are congenital and postoperative obstruction. Intravenous antibiotic prophylaxis is administered to reduce the risk of septicemia.

POSITIONING AND ANESTHESIA

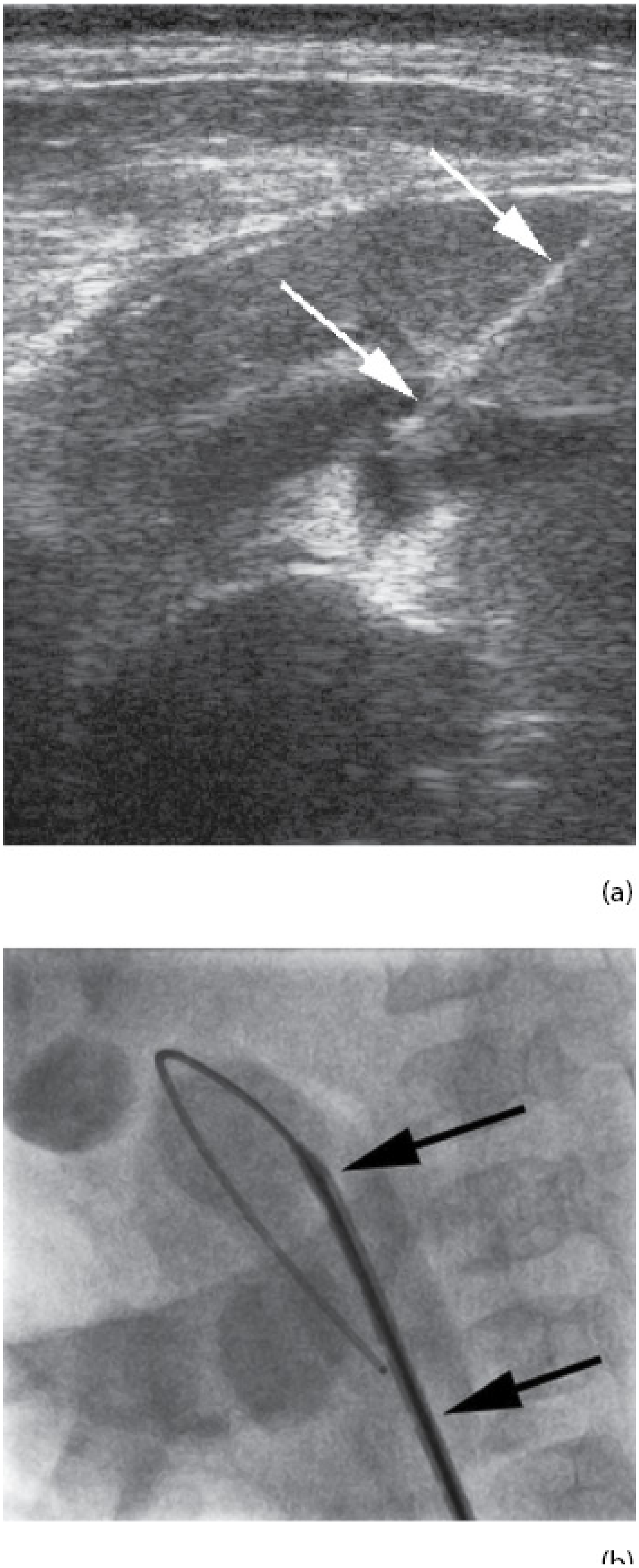
Unilateral nephrostomy is best performed with the patient in a semi-prone position, with the affected side tilted up about 20–30 degrees. This facilitates puncture of a posterior lower-pole calyx through the relatively avascular zone of Brödel. We prefer to perform bilateral nephrostomy under GA with endotracheal intubation, with the patient prone. This requires angling the needle at about 20–30 degrees to the tabletop to achieve the same trajectory (**Figure 90.8**). In addition, the tip of the needle is angled cranially, to make it easier to see at US. This technique also facilitates puncture of an interpolar or upper-pole calyx, which is sometimes required for ureteral interventions or stone removal.

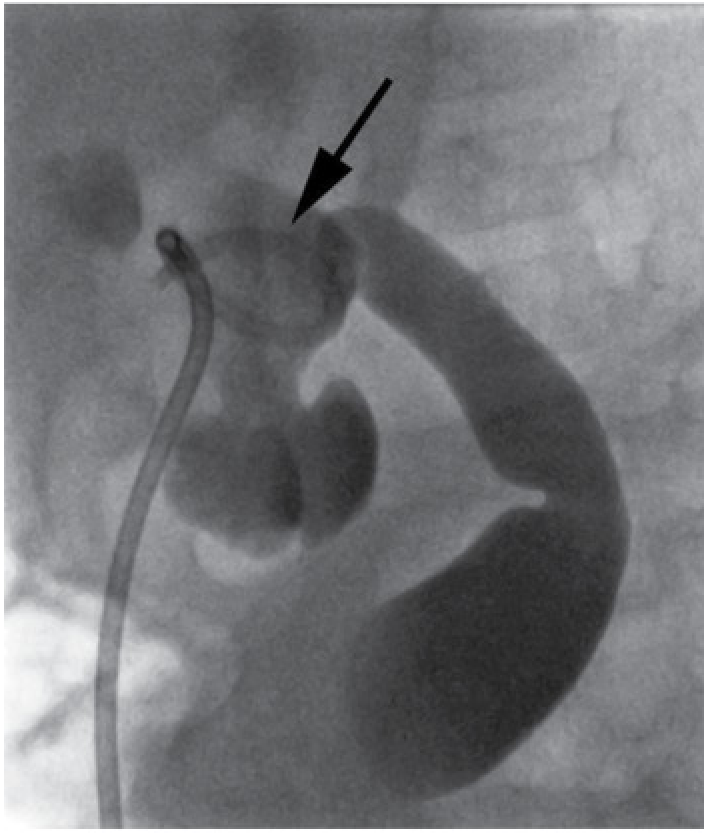


90.8

TECHNIQUE

The exit site and proposed needle tract should be injected with local anesthetic. Using real-time US guidance, a dilated calyx can almost always be punctured with a single needle pass. Various types of needle are available for this purpose. Trocar or sheathed needles have the advantage that, when the trocar is removed, the outer part of the needle (or sheath) has a blunt tip, which is unlikely to damage the collecting system. Needles with echogenic tips are easier to see at US, but they are rarely necessary in children. Once the calyx has been punctured, urine is aspirated for culture, and a smaller volume of dilute contrast (to avoid overdistension of the pelvicalyceal system) is then injected to perform antegrade pyelography (**Figure 90.9a–c**).


(a)
(b)



(c)

90.9a–c

In general, 5 Fr or 6 Fr locking pigtail catheters are appropriate for neonates. In older children, and those with suspected pyonephrosis, larger catheters should be used (typically 8.5 Fr). The use of a stiff wire with a floppy tip is recommended. The largest diameter guidewire that can pass through the stiffener of the pigtail catheter is selected, and the smallest needle that accepts the guidewire is used for the puncture. Once the tip of the guidewire is coiled in the renal pelvis, the tract can be overdilated by 0.5–1 Fr before inserting the pigtail catheter and stiffener. The author usually sutures the catheter to the skin. Coaxial access sets, which allow puncture with a 21- or 22-gauge needle, are more difficult to use, but may be helpful when the pelvicalyceal system is not dilated. With these systems, a 0.018-inch (0.46 mm) guidewire is inserted through the thin needle, and is then exchanged for a 0.035-inch (0.89 mm) wire after insertion of a two-part coaxial dilator.

COMPLICATIONS

The most important complication is injury to an intrarenal branch of the renal artery. This may require embolization (see below). Injuries to the colon and hemothorax or pneumothorax are unusual.

Ureteral dilation and antegrade stenting

Ureteral dilation is most often required in children who have transplant ureteral strictures. Antegrade access is performed as for nephrostomy, but preferably using a mid-pole (or if possible an upper-pole) calyx. The renal pelvis and ureter are opacified with dilute contrast. A peel-away sheath is inserted and the stricture is crossed with a hydrophilic guidewire and a catheter of appropriate shape. With the tip of the guidewire in the bladder, a balloon dilation catheter is positioned across the stricture and inflated with fluoroscopic guidance. A small diameter balloon (3–5 mm, depending on the size of the child) is usually adequate. A cutting balloon may be used if there is a persistent waist.

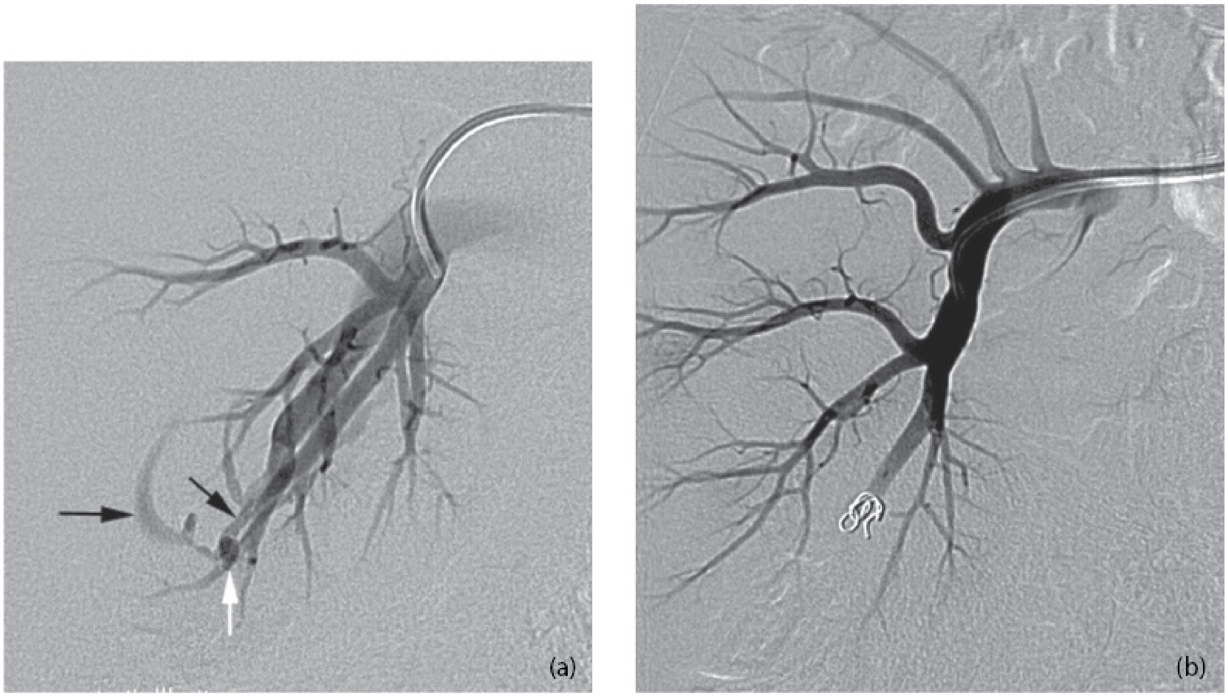
A double-J stent can be placed in the ureter by advancing it over a guidewire with a pusher catheter. A protective nephrostomy tube is probably not necessary unless there is significant bleeding. Occasionally, transurethral removal of a double-J stent is difficult, for example when the lower end of the stent migrates up into the ureter. The stent can then be snared and removed percutaneously, after puncture of the pelvicalyceal system.

BILIARY INTERVENTION

Diagnostic percutaneous transhepatic cholangiography (PTC) may be useful in certain children with suspected biliary strictures, stenosis, or sclerosing cholangitis related to ulcerative colitis or Langerhans' cell histiocytosis. PTC is usually performed at the same time as liver biopsy. When the intrahepatic bile ducts are not dilated, the simplest method is to puncture the gall bladder under US guidance. PTC, with or without biliary drainage or balloon dilation of biliary strictures, may be required following liver transplantation. Malignant obstructive jaundice is unusual in children but biliary drainage with an external or internal–external drainage catheter may be required in some patients.

VASCULAR INTERVENTION

Diagnostic arteriography and venography are required much less frequently now than in the past, but interventional angiography is increasing in importance (**Figure 90.10a and b**). Arterial embolization is an effective method of controlling iatrogenic or other hemorrhage. An example is symptomatic arteriovenous or arteriocalyceal fistula following renal biopsy. These lesions may be embolized with metal coils, which are easy to use and very effective. The aim is to preserve as much renal tissue as possible.



90.10a,b

Arterial embolization may also be used for the treatment of arteriovenous malformations and tumors. The management of vascular malformations (see Chapter 85) and arterial interventions, such as angioplasty and stenting, are beyond the scope of this chapter.

TRACHEOBRONCHIAL INTERVENTION

These techniques are based on bronchography. Balloon dilation of acquired tracheobronchial stenoses is easy and effective. The procedure is analogous to esophageal dilation. The airway can be accessed through an endotracheal or tracheostomy tube, or a laryngeal mask airway. The measurement facilities on modern angiographic equipment are helpful because accurate assessment of the diameter and length of the balloon to be used is very important. Fluoroscopic guidance is useful because it allows accurate positioning of the balloon, and abolition of the waist confirms that complete inflation has been achieved. A description of stenting and other pediatric airway intervention is beyond the scope of this chapter.

FURTHER READING

Barnacle AM, Wilkinson AG, Roebuck DJ. Paediatric interventional uroradiology. *Cardiovasc Intervent Radiol* 2011; 34: 227–40.

Hogan MJ, Hoffer FA. Biopsy and drainage techniques in children. *Tech Vasc Interv Radiol* 2010; 13: 206–13.

Lord DJ. The practice of pediatric interventional radiology. *Tech Vasc Interv Radiol* 2011; 14: 2–7.

Marshalleck F. Pediatric arterial interventions. *Tech Vasc Interv Radiol* 2010; 13: 238–43.

Roebuck DJ, Hogan MJ, Connolly B, McLaren CA. Interventions in the chest in children. *Techn Vasc Interv Radiol* 2011; 14: 8–15.

Roebuck DJ, McLaren CA. Gastrointestinal intervention in children. *Pediatr Radiol* 2011; 41: 27–41.

91

Exposure for spinal surgery

STEVEN W. BRUCH

PRINCIPLES AND JUSTIFICATION

Safe exposure of the anterior portion of the spine allows the orthopedic surgeon to resect, manipulate, or instrument the spine to treat various abnormalities. The pediatric surgeon moves important structures out of the way to provide adequate and safe exposure for the orthopedic surgeons.

INDICATIONS

The orthopedic indications for procedures requiring anterior spinal exposure are varied ranging from deformities (scoliosis and kyphosis), to mass resections, to traumatic injuries. The pediatric surgeons rely on their orthopedic colleagues to ensure the surgical indications are sound, but should review the imaging studies and have a clear plan with the orthopedic surgeon on the degree of exposure required.

PREOPERATIVE ASSESSMENT

The preoperative evaluation should concentrate on comorbid disease, and the neurologic and peripheral vascular examinations. These procedures often involve manipulation of the spine and major vessels including the aorta, inferior vena cava (IVC), and their branches, so a baseline vascular and neurologic examination is key. It is especially important to obtain preoperative renal function studies since correction of the spinal deformity changes the contour of the aorta and its relationship with the renal artery, which can affect renal blood flow and even cause renal failure postoperatively. In addition, acute changes in the curvature of the spine may compress the spinal cord or nerve roots with compromise in nerve function. The chest, abdomen, and flanks should also be evaluated for evidence of previous surgical procedures as they may impact the placement of the incision.

ANESTHESIA

General anesthesia is required with endotracheal intubation. The degree of monitoring depends on comorbid diseases, and ranges from peripheral IV lines to central venous and arterial lines. Transcranial motor evoked potential and somatosensory evoked potential (SSEP) monitoring of the posterior tibial nerve, with the ulnar nerve as a control, is commonly utilized.

OPERATION

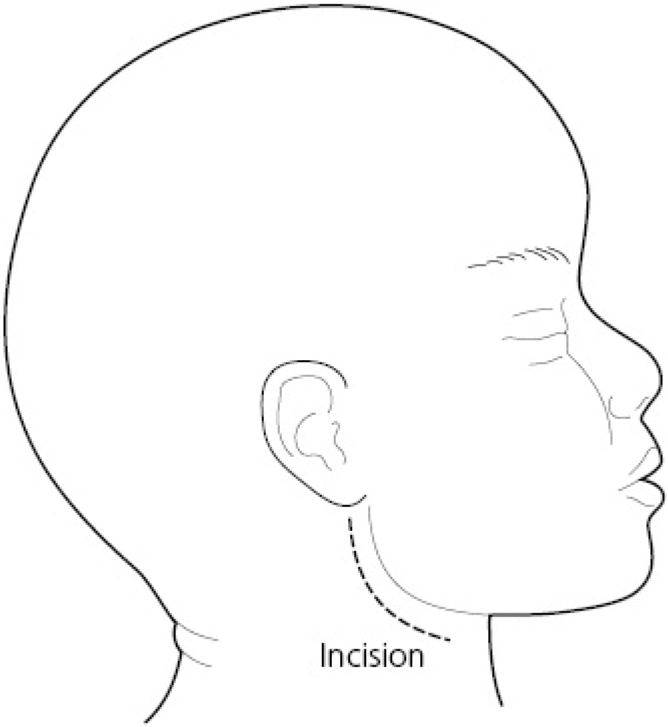
Several approaches are used depending on the target of the orthopedic or neurosurgeon.

C1–C2

A transoral approach allows access to the anterior aspect of C1 and C2. With the neck extended, a Dingmann's clamp is placed in the mouth, and a midline incision opens the posterior pharyngeal wall. Remaining in the midline allows separation of the longus capitis and longus colli muscles, exposing the anterior longitudinal ligament, which is divided in the midline. No neurovascular structures should be encountered by staying midline, thus avoiding injury. The vertebral artery is located about 1 cm lateral to the midline along with the vertebral venous plexus which is difficult to control if entered.

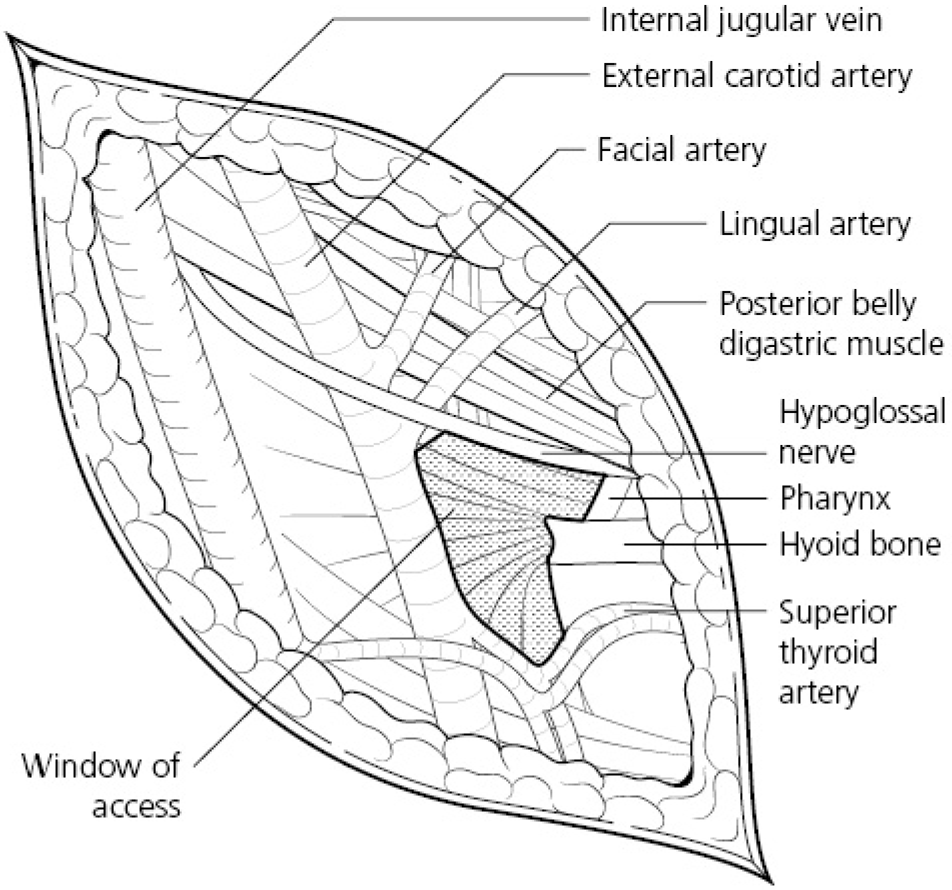
C3

The approach to the anterior aspect of C3 lies between the carotid sheath, which is reflected laterally, and the cervical visceral column, which is reflected medially. The pathology determines the laterality of the approach. The neck is extended and turned slightly to the contralateral side. An incision is made from the mastoid process, passing under the angle of the mandible and continuing anteriorly inferior to the mandible (**Figure 91.1**). The platysma is incised in the same direction as the skin incision. The cutaneous sensory branches of the cervical plexus (lesser occipital, greater auricular, and transverse cervical nerves) should be avoided.



91.1

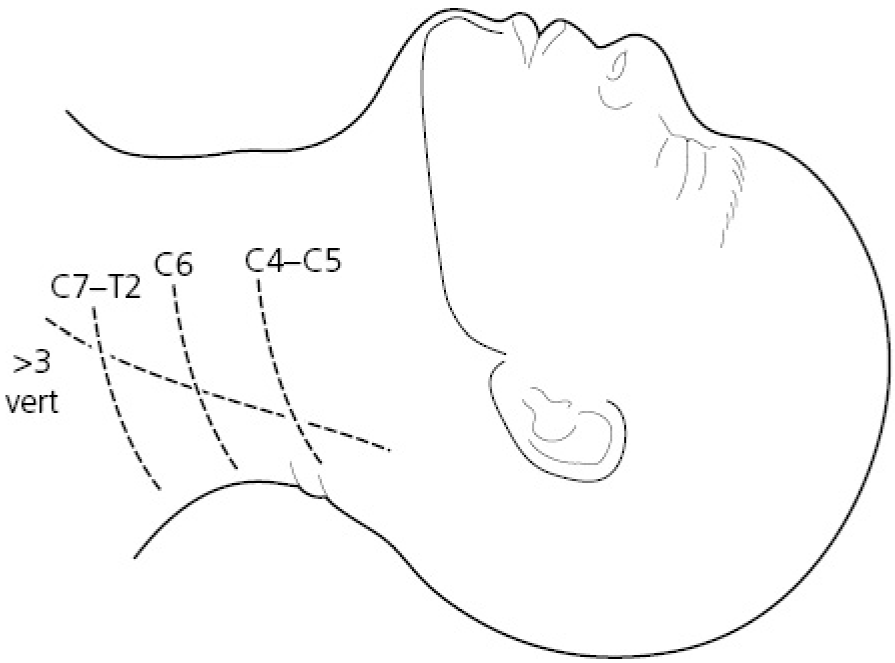
The sternocleidomastoid muscle (SCM) is then retracted laterally and the "window of access" is opened. This window is bounded by the external carotid artery laterally, the pharynx at the level of the hyoid bone medially, the hypoglossal nerve superiorly, and the superior thyroid artery inferiorly (**Figure 91.2**). As the pharynx is mobilized medially, the anterior surface of the vertebral column is reached. A nasogastric tube or bougie in the esophagus helps avoid injury during mobilization. The longus colli and longus capitus are separated in the midline, and the prevertebral fascia and anterior longitudinal ligament are divided in the midline to expose the anterior aspect of C3.


Internal jugular vein
External carotid artery
Facial artery
Lingual artery
Posterior belly digastric muscle
Hypoglossal nerve
Pharynx
Hyoid bone
Superior thyroid artery
Window of access

91.2

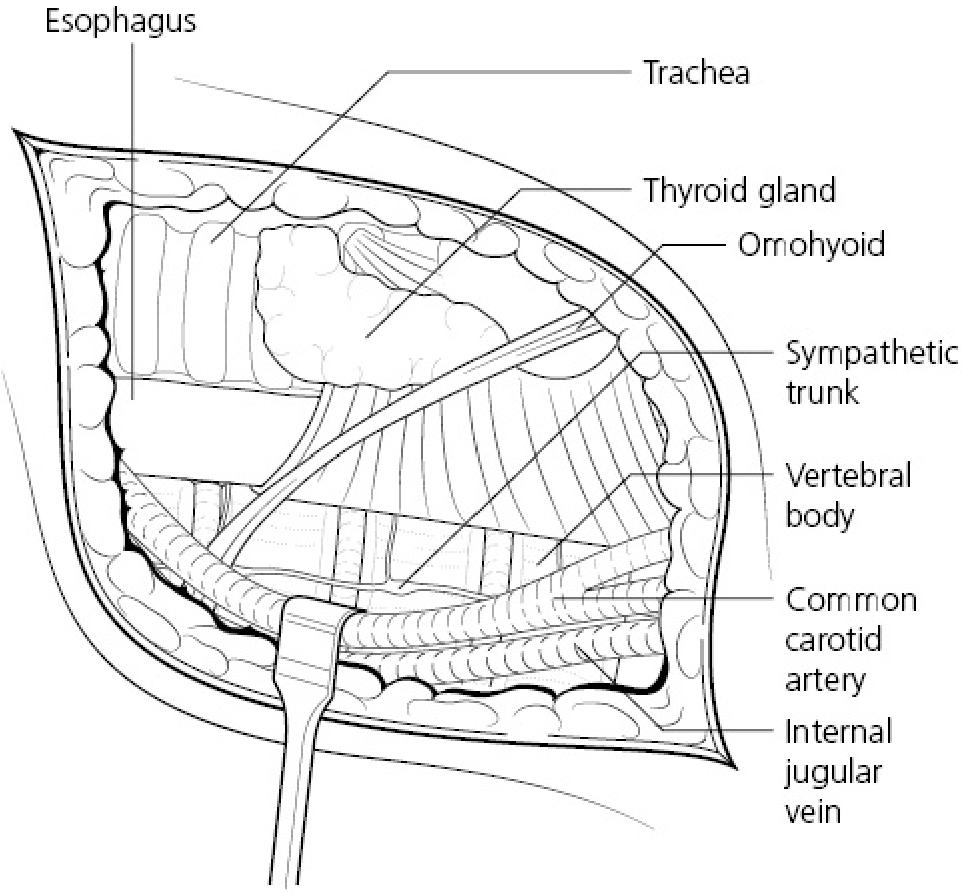
C4–T2

Several maneuvers provide exposure of the anterior surface of the mid to lower cervical vertebrae. The most useful is the anteromedial approach, which can be extended up to the level of C1, if necessary. Either side of the neck can be used with this approach, but the left side is preferable for access to vertebral levels C6–T2 to help avoid the recurrent laryngeal nerve, which lies more lateral on the right side. The neck is extended with the head turned to the contralateral side, opening the space between the neurovascular bundle and the cervical viscera. The skin incision is centered on the anterior aspect of the SCM and placed in the skin lines. The position of the incision depends on the vertebral level of interest. The incision to approach C4–C5 begins at the upper aspect of the thyroid cartilage, for C6 the incision begins at the cricoid cartilage, and for C7–T2 the incision begins a finger's breadth above the sternal notch. An incision along the anterior border of the SCM can be used if multiple levels require exposure (**Figure 91.3**).



91.3

The platysma is divided in the direction of its fibers, and the deep cervical fascia is incised just medial to the SCM. The strap muscles along with the cervical viscera (thyroid gland, larynx, trachea, pharynx, and esophagus) are brought medial, and the neurovascular bundle is retracted laterally to expose the vertebral column (**Figure 91.4**). The hypoglossal and recurrent laryngeal nerves must be avoided during this maneuver. Identification of the esophagus improves with the insertion of a nasogastric tube or bougie, as the decompressed esophagus is a flat structure making it difficult to identify and protect from injury. The anterior longitudinal ligament is then opened in the midline at the appropriate level. The prevertebral musculature can be detached laterally, but only for about 1 cm to avoid injury to the vertebral vasculature and the sympathetic trunk.

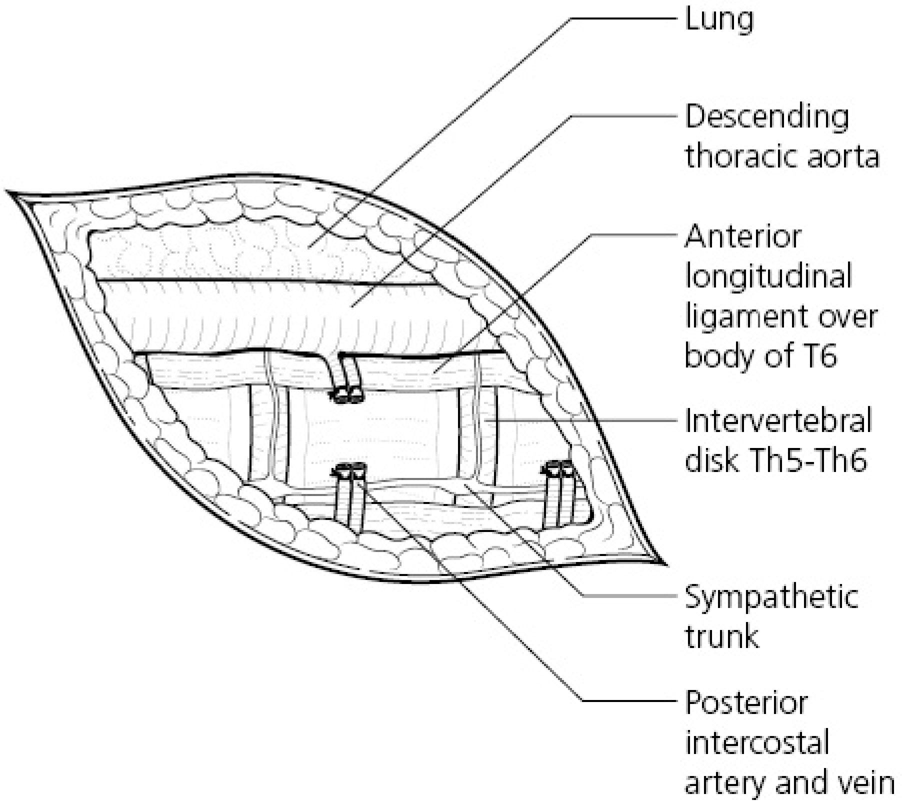

Esophagus
Trachea
Thyroid gland
Omohyoid
Sympathetic trunk
Vertebral body
Common carotid artery
Internal jugular vein

91.4

T3–T10: Open operation

A posterolateral thoracotomy allows access to the anterior vertebral bodies from T3 to T10. The pathology dictates the side of the thoracotomy. For kyphoscoliotic disease, in general, the incision is made on the side of the apex of the curve. Since the aorta provides a bit of a challenge on the left, a left-sided approach will be described. The skin incision depends on the level of the target. The incision should be made one rib level higher than the target vertebral level. If in doubt, choose the higher rib since exposure inside the chest is much easier to extend inferiorly than superiorly. A muscle-sparing technique should be used when feasible.

The rib that the incision is based on is then resected by incising the periostium and stripping it off the superior aspect of the rib. This is performed using a Freer elevator, then a Doyen elevator to circumferentially dissect the periostium. The rib is then divided at the costochondral junction and removed as far posteriorly as possible. The rib is preserved for the orthopedic surgeons to use as bone grafts later in the procedure. The chest is then entered through the resected rib bed using an appropriately sized Finochetto for retraction. The mediastinal pleura is incised between the descending aorta and the hemiazygos vein. In certain situations, it may be preferable to stay retropleural. The aorta is rolled medially to expose the vertebral body and intervertebral disks (**Figure 91.5**). Segmental vessels give rise to the posterior intercostal vessels and the blood supply to the spinal cord. These vessels should be divided only if necessary, and before they branch and give rise to the anastomotic plexus that feeds the spinal cord. Spinal cord compromise may be predicted prior to vessel division using evoked potentials following the placement of a vascular clamp (bulldog) on the selected artery. With continued mobilization, the anterior longitudinal ligament and vertebra are visualized. The thoracic duct should be avoided. It will come into play on the left side in the area of the third and fourth thoracic vertebra where it crosses the midline just ventral to the vertebral column. The duct is present in the right chest inferiorly at the diaphragmatic crura close to the esophagus, coursing superiorly until it crosses the midline. Following the procedure, the chest is closed with a chest tube remaining in place.

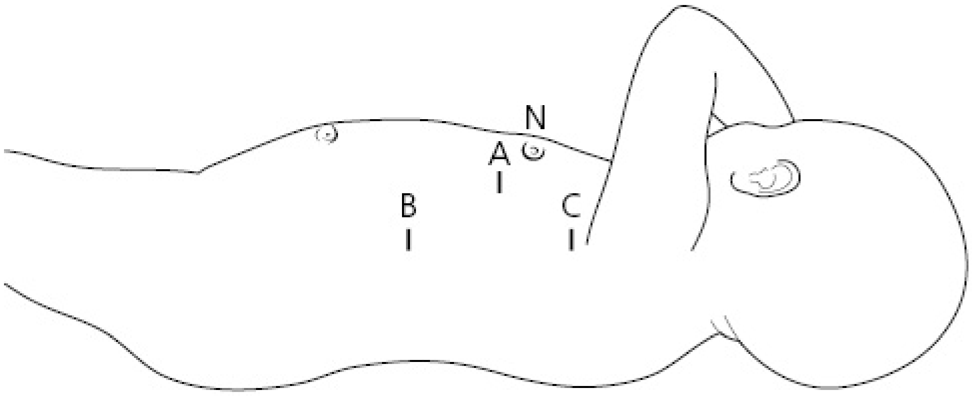

Lung
Descending thoracic aorta
Anterior longitudinal ligament over body of T6
Intervertebral disk Th5-Th6
Sympathetic trunk
Posterior intercostal artery and vein

91.5

T3–T10: Thoracoscopic operation

A thoracoscopic approach to the vertebrae in the chest may be appropriate in cases that do not require significant anterior instrumentation. The approach can be from either side utilizing the lateral decubitus position. Isolation of the ipsilateral lung is imperative for success. The anesthesiologists may use a double-lumen endotracheal tube in larger children, or a bronchial blocker or contralateral mainstem intubation technique in smaller children, and this, combined with gentle insufflation, provides excellent visualization.

The initial trocar is placed after isolation of the ipsilateral lung in the fifth interspace along the anterior axillary line (**Figure 91.6**). The chest is insufflated with CO_2 at up to 5 mmHg pressure, and a 5 mm, 30-degree scope is used. Additional ports and instruments are used to allow dissection for exposure, excision of the disks, and instrumentation, if necessary. The dissection is similar to that in the open description above. The soft tissues are reflected off the anterolateral aspect of the vertebrae after division of segmental vessels if necessary. The segmental vessels are controlled using electrosurgical sealing devices. Following the procedure, the trocars are removed, the lung is re-expanded, and a chest tube is placed through an appropriate trocar site.



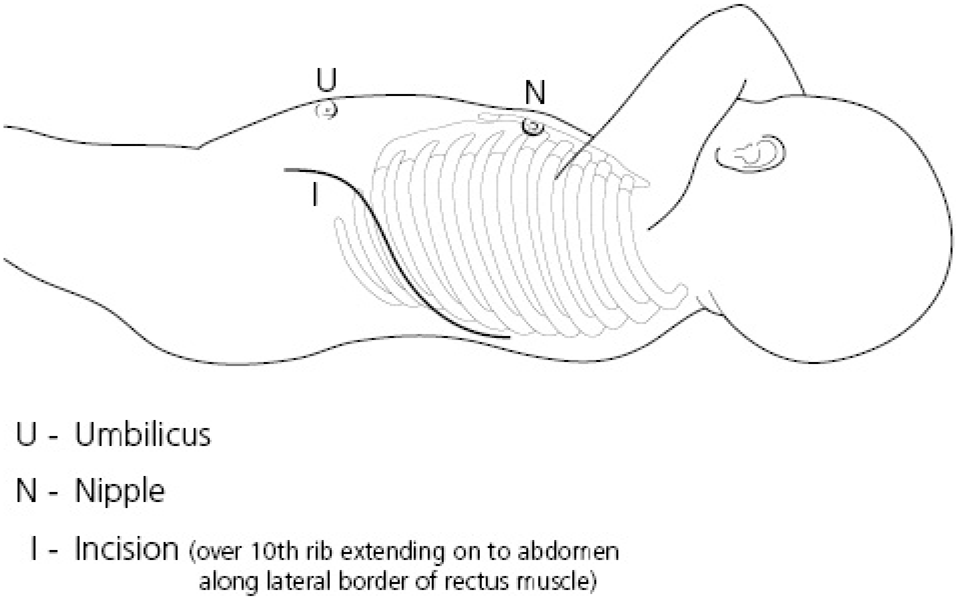
N - Nipple
A - Initial trocar site 5th interspace, anterior axillary line
B - Trocar site - inferior chest in mid axillary line
C - Trocar site - under tip at scapular

91.6

T11–L1

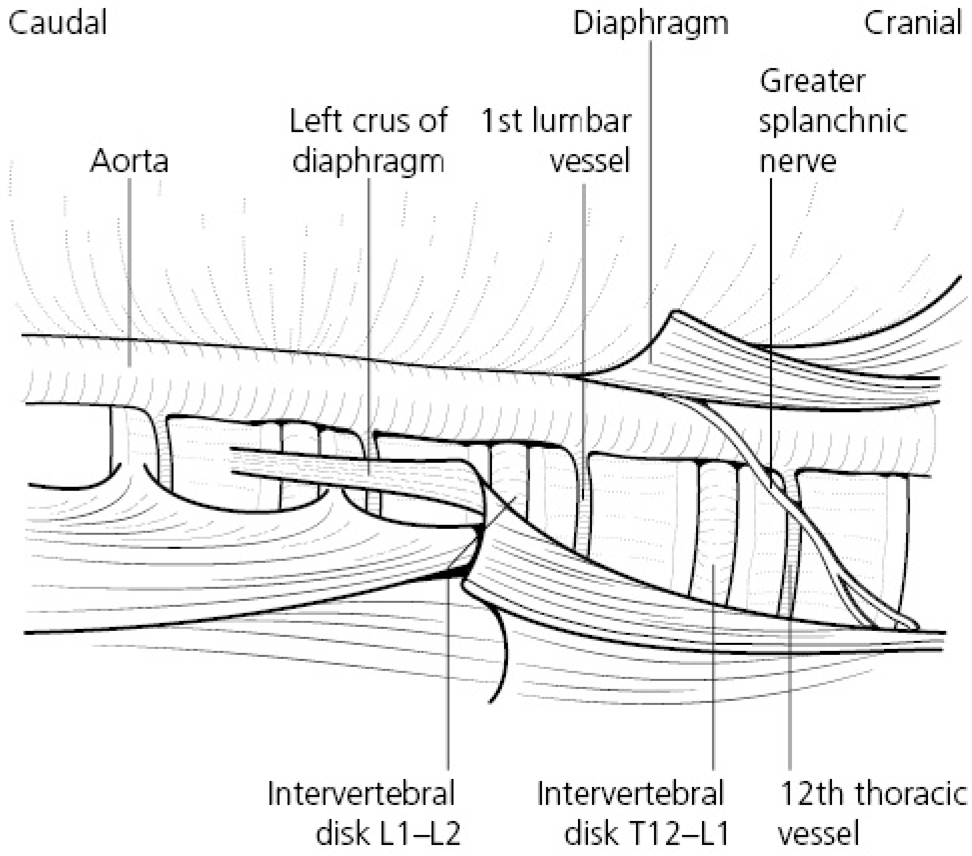
A thoracoabdominal approach through the bed of the tenth rib best exposes the anterior surface of the thoracolumbar junction (T11–L1). The site and type of lesion usually determines the side of the approach, but a left-sided approach would be favored if the lesion does not dictate laterality. The reason for the left-sided approach is that the IVC is riskier to mobilize than the aorta, and the liver is more difficult to retract than the spleen. The lateral decubitus position with the shoulder rotated back toward the bed allows excellent exposure for both the chest and abdominal portions of the procedure.

The skin incision is made along the tenth rib across the costal margin and then inferiorly about 2 cm lateral to the border of the rectus abdominis muscle (**Figure 91.7**). The inferior aspect of the incision is determined by the inferior vertebral level required (the body of L2 lies just cranial to the umbilicus). The incision is taken down to the tenth rib, incising the latissimus dorsi and external oblique muscles. The tenth rib is removed by incising the periosteum and stripping it off the superior aspect of the rib. The rib is then divided at the costochondral junction, elevated, dissected off the neurovascular bundle inferiorly, and divided posteriorly. The rib is saved for bone grafting later. The bed of the tenth rib is opened to gain access to the pleural cavity. The retroperitoneal space is entered by cutting the cartilage of the tenth rib and the underlying diaphragm and transversus abdominis musculature. This allows visualization of the peritoneum and the fatty tissue associated with it.


U
N
I
U - Umbilicus
N - Nipple
I - Incision (over 10th rib extending on to abdomen along lateral border of rectus muscle)

91.7

The diaphragm is opened in a radial fashion, leaving a 2 cm edge to allow adequate reconstruction. Sutures are placed on each side of the diaphragm incision at intervals to allow accurate realignment of the muscle. While dividing the diaphragm, the peritoneum is reflected anteriorly and inferiorly off the diaphragm and the lateral and posterior abdominal walls. To gain access to the vertebral column, the mediastinal pleura is opened just lateral to the thoracic aorta, and the left diaphragmatic crus is divided (**Figure 91.8**). The aorta then must be mobilized to the right to gain access to the anterior longitudinal ligament and the anterior surface of the vertebral column. If division of segmental vessels is required, they should be divided next to the aortic wall to preserve spinal blood flow. The thoracic duct passes behind the aorta at this level and should be avoided.

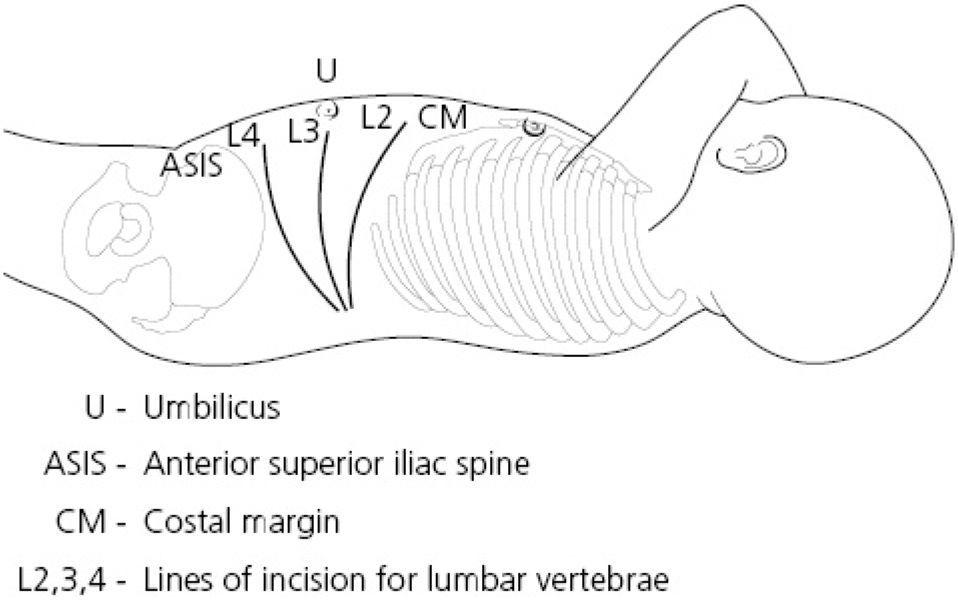

Caudal
Diaphragm
Cranial
Greater splanchnic nerve
Left crus of diaphragm
1st lumbar vessel
Aorta
Intervertebral disk L1–L2
Intervertebral disk T12–L1
12th thoracic vessel

91.8

L2–L4

The lateral extraperitoneal approach allows excellent exposure to the mid-lumbar vertebral column. Again, a left-sided approach is preferable, but the pathology will determine the side of the exposure. A semi-lateral position with the table flexed, using the kidney rest between the iliac crest and costal margin, allows excellent exposure with the surgeon standing in front of the patient.

The skin incision starts at the mid-point between the costal margin and the iliac crest at the mid-axillary line, extends toward the umbilicus, and ends at different points along the edge of the rectus muscle depending on the target level: L2, just superior to the umbilicus; L3, at the level of the umbilicus; and L4, just inferior to the umbilicus (**Figure 91.9**). There are three muscle layers to divide before getting to the level of the peritoneum. The external oblique, internal oblique, and transversus abdominis muscle layers should all be bluntly separated in the direction of their fibers to gain access to the peritoneum. A large blunt hemostat is useful for splitting the muscle layers without entering the peritoneum.

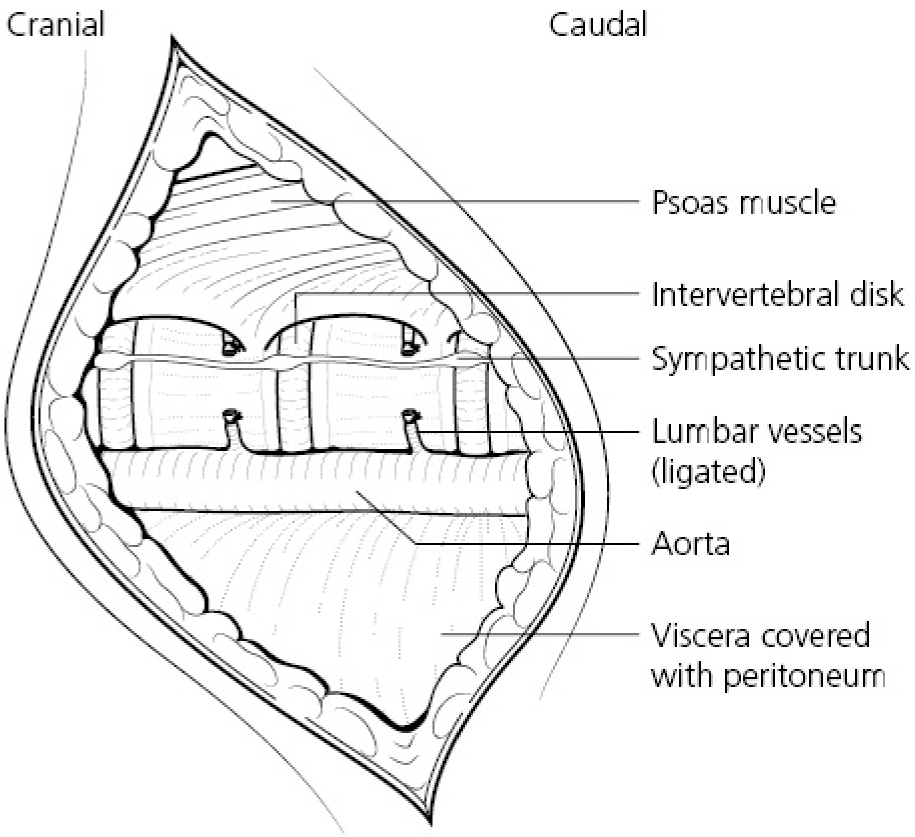

U
L4
L3
L2
CM
ASIS
U - Umbilicus
ASIS - Anterior superior iliac spine
CM - Costal margin
L2,3,4 - Lines of incision for lumbar vertebrae

91.9

An alternative approach creates a longitudinal incision from the costal margin to the anterior superior iliac spine. After the fascia of the external oblique has been divided, the internal oblique and transversalis muscles are bluntly opened to gain access to the retroperitoneal space.

The anterior dissection of the peritoneum should begin as far posterior as possible where there is more extraperitoneal fat making this dissection easier and less likely to enter the peritoneal cavity. If the peritoneum is entered, it should be immediately closed to minimize the extent of the hole.

A hole in the peritoneum allows the extruded bowel to significantly compromise the exposure. The tissues anterior to the psoas muscle, including the ureter, are mobilized medially to gain access to the vertebral column. The aorta on the left and the IVC on the right are then mobilized medially to expose the lumbar vertebrae. There are lumbar arteries and veins that tether the aorta and IVC to the vertebral column and require division to gain exposure to the anterior aspect of the vertebral column (**Figure 91.10**). At the L5 area, the segmental veins enter the common or internal iliac vein on the left, called the iliolumbar vein. If this is not controlled prior to mobilization of the vessels, it can easily tear and be difficult to control.

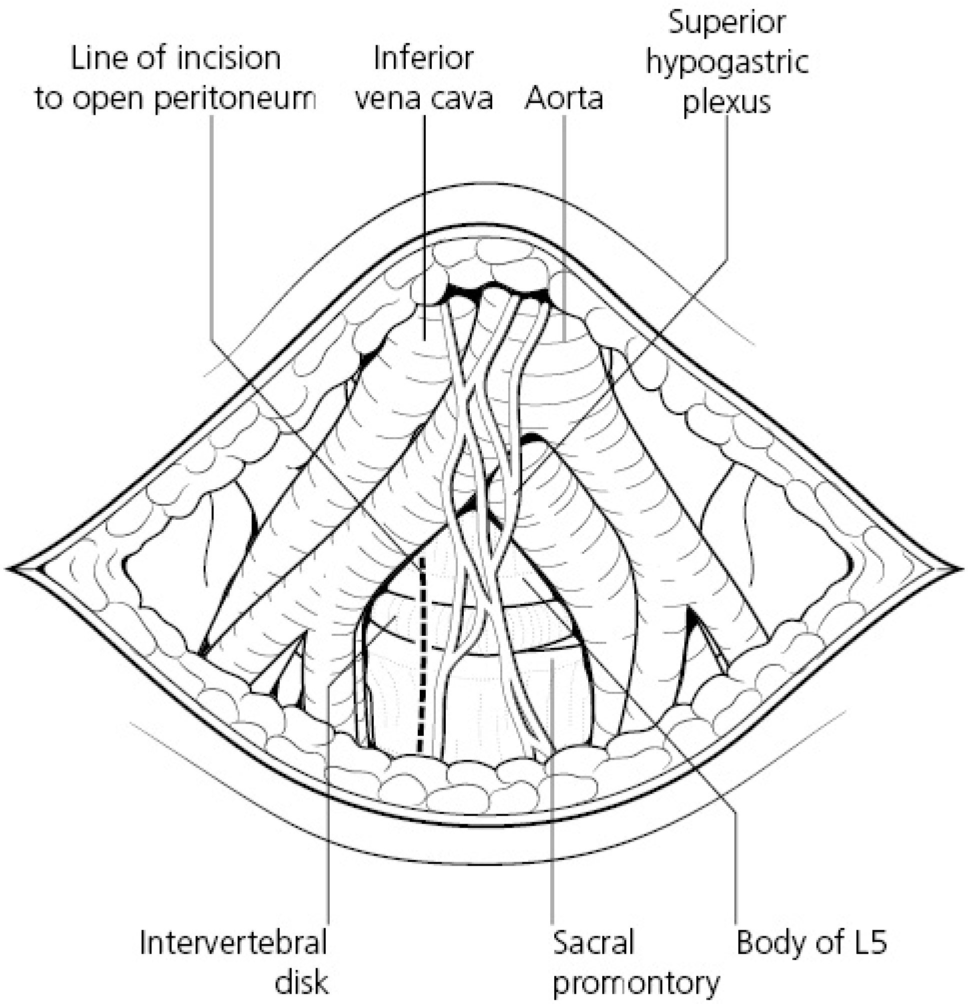

Cranial
Caudal
Psoas muscle
Intervertebral disk
Sympathetic trunk
Lumbar vessels (ligated)
Aorta
Viscera covered with peritoneum

91.10

The dissection is completed by sweeping off the remainder of the prevertebral connective tissue, which may lead to bleeding from the prevertebral venous plexus. The L5 region may be challenging due to take-off of the iliac vessels, and an anterior approach (see below) may be more optimal.

L5–S1

A transperitoneal approach provides the best exposure for the lumbosacral junction. In the supine position, a Pfannenstiel incision with a midline opening from the symphysis to the umbilicus provides the required exposure. A Foley catheter decompresses the bladder. With the help of Trendelenburg positioning, the small intestine is mobilized superiorly and the sigmoid colon to the left, allowing visualization of the sacral promontory and the aortic and IVC bifurcations (**Figure 91.11**). A self-retaining retractor facilitates exposure.


Line of incision
to open peritoneum
Inferior
vena cava
Aorta
Superior
hypogastric
plexus
Intervertebral
disk
Sacral
promontory
Body of L5

91.11

The middle sacral vessels require division. The iliac arteries and veins may or may not require mobilization depending on their location. The iliolumbar vein must be divided if significant mobilization of the left iliac vein is required, to prevent troublesome bleeding if it is avulsed. The superior hypogastric plexus of nerves, an autonomic nerve plexus that controls the ejaculatory mechanism, lies distal to the inferior mesenteric artery over the aorta and extends over the left common iliac vein and L5, and L5–S1. At the level of S1, it divides into the right and left hypogastric nerves. It lies close to the midline, but often somewhat to the left of it. To preserve this nerve plexus, the posterior peritoneum overlying the L5–S1 area should be opened to the right of midline medial to the ureter, and the medial tissue should be bluntly (without the use of electrocautery) dissected to the left to lift the superior hypogastric nerve plexus off the vertebra and preserve it while gaining access to the anterior aspect of the lumbosacral junction.

POSTOPERATIVE CARE

Postoperative care differs depending on the location of the exposure and is shared with the orthopedic surgeon. The exposures of C1–T2 involve transoral or neck incisions. These patients are often left intubated overnight and observed for bleeding and airway issues. Their diets can then be advanced following extubation. The exposures of T3–L1 involve a thoracotomy, thoracoabdominal, or thoracoscopic approach, all of which require chest tube drainage. The chest tube can be removed if there is no air leak or significant fluid output (less than 1 mL/kg per 8-hour shift). Early removal of chest tubes is especially important when anterior hardware has been placed to reduce the risk of infection. The diet can be advanced on the first postoperative day. The patients who require a thoracoabdominal approach may experience an early postoperative ileus, especially if the duodenum is mobilized, and should have their diets advanced accordingly. The exposures of L2–S1 require retroperitoneal mobilization or a transperitoneal approach and may induce a postoperative ileus. The diet is advanced as the ileus resolves. The thoracoabdominal cases and children with severe comorbid illness often require an initial postoperative stay in the intensive care unit.

COMPLICATIONS

Intraoperative

Intraoperative complications vary depending on the level of the exposure. The majority of intraoperative complications relate to vessel injury. Paraplegia is a concern, especially when the exposure requires the division of multiple segmental arteries to the spine. The mid-thoracic spine (T4–T9) is at greatest risk for ischemia leading to paralysis. If these segmental vessels require division, it should take place as close to the aorta as possible, and be limited to one side of the spine to retain the collateral circulation. Test occlusion with SSEP monitoring should guide division. Injuries to nerves can occur in different areas. The recurrent laryngeal nerve can be injured directly or secondary to a stretch injury from retraction when exposing the cervical and upper thoracic spine. The hypogastric plexus of nerves must be avoided when exposing the lower lumbar and sacral spinal levels. Injury to this plexus can result in retrograde ejaculation. An injury to the thoracic duct can result in a chylothorax.

Postoperative

The majority of postoperative complications are related to the respiratory system. A review of 505 children who had an anterior spinal exposure reported a 9.8% overall complication rate with half of the complications being respiratory in nature, including effusions, pneumothoraces, atelectasis, and respiratory failure. Gastrointestinal complications included superior mesenteric artery syndrome, which resolved with conservative management (parenteral nutrition or use of a nasojejunal feeding tube), and gastrointestinal bleeding. The other complications reported in this review were vessel injuries at the time of the exposure and urinary tract infections. A more recent review of over 19 000 children with scoliosis surgery revealed fewer but similar complications, with a 1% incidence of neurologic and pulmonary complications, and a 2.3% risk of a wound infection.

FURTHER READING

Crawford BK, Morgan JA. Thoracic exposures for spinal deformity surgery. In: Errico TJ (ed.). *Surgical Management of Spinal Deformities*, 1st ed. Philadelphia: Saunders; 2009: pp. 503–10.

Fasel JHD. *Anterior Approaches to the Vertebral Column.* Toronto: Hogrefe & Huber; 1991: pp. 11–49.

Gardocki RJ. Spinal anatomy and surgical approaches. In: Canale ST, Beaty JH (eds). *Campbell's Operative Orthopedics*, 11th ed. St Louis: Mosby; 2007: pp. 1735–51.

Janik JS, Burrington JD, Janik JE, et al. Anterior exposure of spinal deformities and tumors: A 20-year experience. *J Pediatr Surg* 1997; 32: 852–9.

Reames DL, Smith JS, Fu K-MG, et al. Complications in the surgical treatment of 19,360 cases of pediatric scoliosis. *Spine* 2011; 36: 1484–91.

92

Surgery of conjoined twins

LEWIS SPITZ, EDWARD M. KIELY, AND AGOSTINO PIERRO

HISTORICAL HIGHLIGHTS

The earliest example of conjoined twins is a 17 cm marble statuette portraying parapagus twins, "the double goddess," dating from the sixth millennium BCE. The statue of the sisters of Catathoyuk is in the collection of the Anatolian Civilization Museum in Ankara, Turkey. Another early example is a stone carving of pygopagus twins dated to 80 BC in the St Marco Museum, Florence, Italy. The earliest attempt at separation of conjoined twins occurred in Kappadokia, Armenia, in AD 970. When one of the male ischiopagus twins died, aged 30 years, an attempt was made to save the surviving twin by separating him from his dead brother, but he died 3 days later.

The first well-documented case is that of the Biddenden maids born in Kent, England, in AD 1100 who were reputed to have been joined at the hips and shoulders. They lived together for 34 years. When Mary fell ill and died, Eliza was advised to be separated from her dead sister but absolutely refused, saying, "As we came together we will also go together." She died 6 hours later.

The first successful separation of conjoined twins took place in 1689. The surgeon, Johannes Fatio, separated omphalopagus twins in Basel, Switzerland, by "tracing the umbilical vessels to the navel where he tied them separately. He then transfixed and tied the bridge between the two infants with a silken cord." The ligature fell off on the ninth postoperative day and both children survived.

The most celebrated pair of conjoined twins was Chang and Eng Bunker, born on a river boat on the Mekong River, Siam, in 1811. They were joined at the xiphisternum by a short bridge that stretched so they were eventually able to stand side by side. They were taken to the United States where they were widely exhibited by the showman Phineas Barnum. They married sisters, lived in North Carolina, and had 21 children between them. They lived together for 62 years.

INCIDENCE AND ETIOLOGY

The frequency of conjoined twins has been estimated at 1 in 250 000 live births. Sixty percent of conjoined twins die during gestation or at birth. Females predominate with a ratio of 3 : 1.

The most widely held theory of their occurrence is that secondary fusion occurs between two originally separate monovular embryonic disks at around 13–15 days' gestation ("fusion" theory of Spencer). An alternative theory is that of failure of complete separation of the embryonic disk ("fission" theory).

CLASSIFICATION

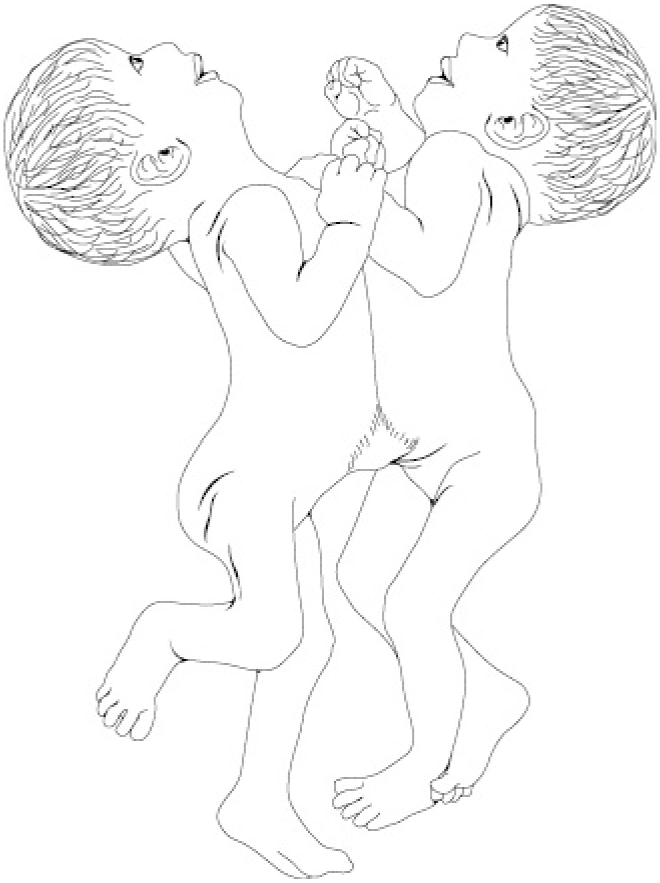
Conjoined twins are classified on the basis of the site of union, with the suffix "pagus" meaning fixed or fastened (**Table 92.1**). The twins can have four (tetrapus), three (tripus), or two (bipus) legs.

Table 92.1 Classification of conjoined twins

	Percent occurrence (%)	
Classification	**Literature**	**Great Ormond Street series**
Thoracopagus	40	12
Omphalopagus	33	9
Pygopagus	19	4
Ischiopagus	6	5
Craniopagus	2	3
Parapagus	–	5

Thoracopagus

The twins lie face to face (**Figure 92.1**) and share the sternum, diaphragm, upper abdomen wall, and liver and have an exomphalos.



92.1 Thoracopagus.

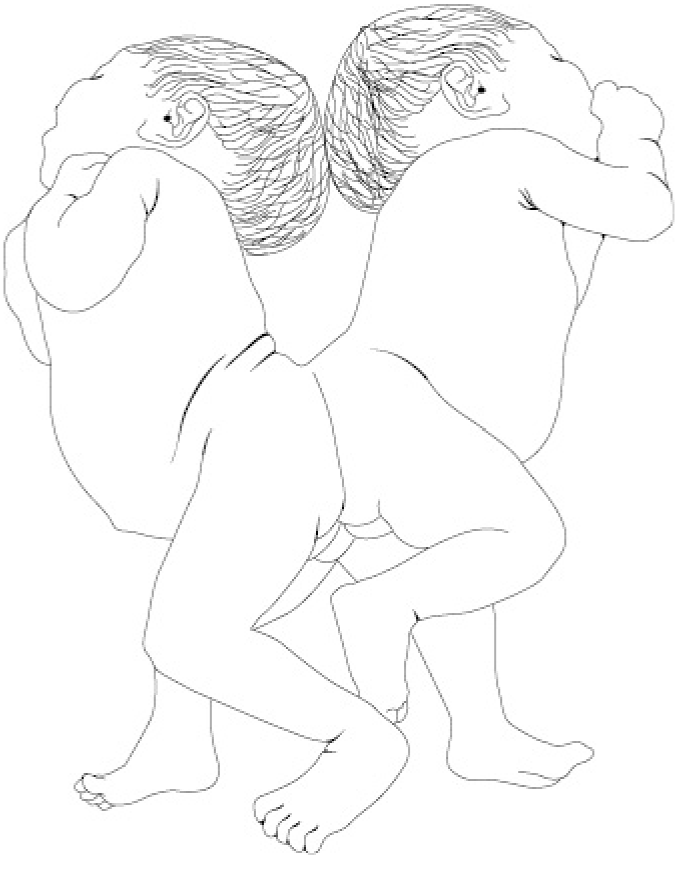
In the majority of cases, they share the pericardium (90%) and heart (85%). They may have a common small intestine (50%) which joins at the duodenum and separates at the ileum; the biliary tree may be joined in 25% of patients. The twins may have associated cardiac anomalies, such as ventricular septal defect, atrial septal defect, and tetralogy of Fallot.

Omphalopagus

The twins are joined ventrally in the abdomen, often including the lower thorax. The heart is never fused although the pericardium may be shared. The liver is shared in 80% of cases and there is often an omphalocele. The distal duodenum and small intestine to the level of the Meckel's diverticulum may be shared, but the terminal ileum and colon are separate. The genitourinary (GU) tract is separate.

Pygopagus

The twins are joined dorsally (**Figure 92.2**), sharing the sacrococcygeal and perineal regions. They face away from each other, and share the sacrum, coccyx, and part of the pelvic bones. The spinal cords are usually separate. Twenty-five per cent share the lower GI tract and have a single anus and one or two rectums. In 15% of cases, there is a single bladder. There is an increased incidence of vertebral anomalies, including hemivertebrae, hemisacral agenesis, and thoracic anomalies.



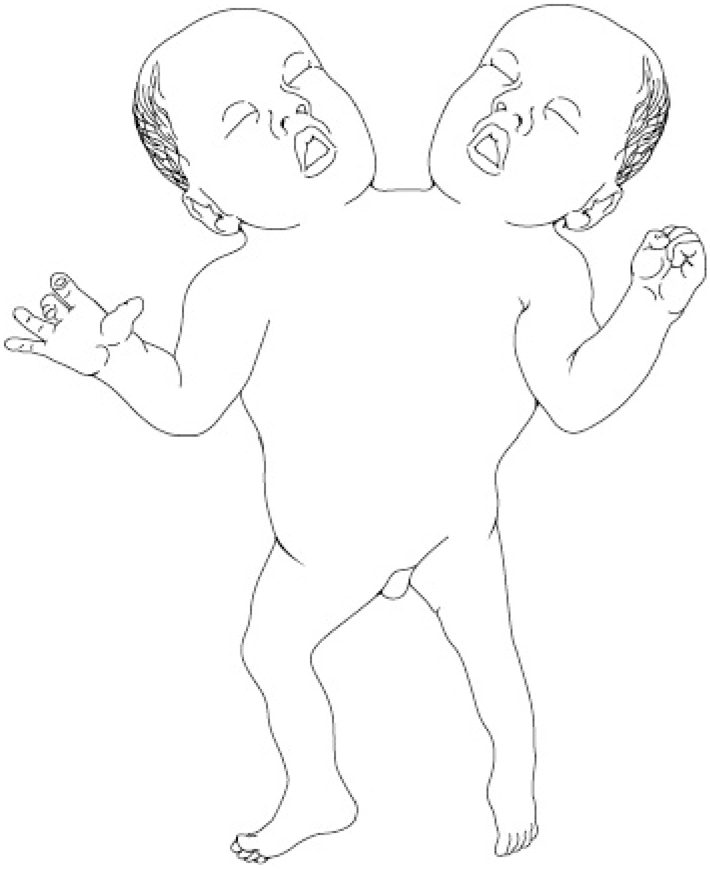
92.2 Pygopagus.

Although the pelvic conjunction is fundamentally different than in ischiopagus twinning, the types are similar insofar as numerous other associated orthopedic anomalies have been reported in association with pelvic conjunction, such as hip subluxation or dislocation, congenital vertical talus, talipes equinovarus, Sprengel shoulder, and scoliosis.

There can also be a variable degree of spinal and cord fusion. Although there may be only one anus and rectum, the remainder of the intestines are usually separate. The upper bodies are not fused and there are four arms and four legs.

Parapagus

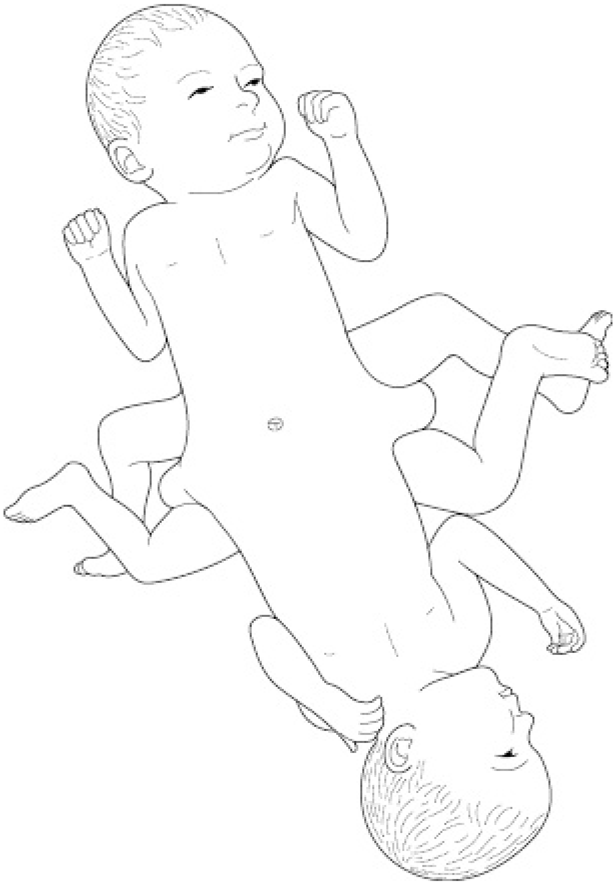
This is a relatively new term denoting extensive side-to-side fusion (**Figure 92.3**). The twins lie side-to-side and share the umbilicus, abdomen, liver, pelvis (single symphysis pubis), and the GU tract. They have a common colon and an anorectal anomally and colovesical fistula or cloaca anomaly. The twins have separate heads, but the entire trunk is conjoined. They may have two, three, or four arms and two or three legs.



92.3 Parapagus.

Ischiopagus

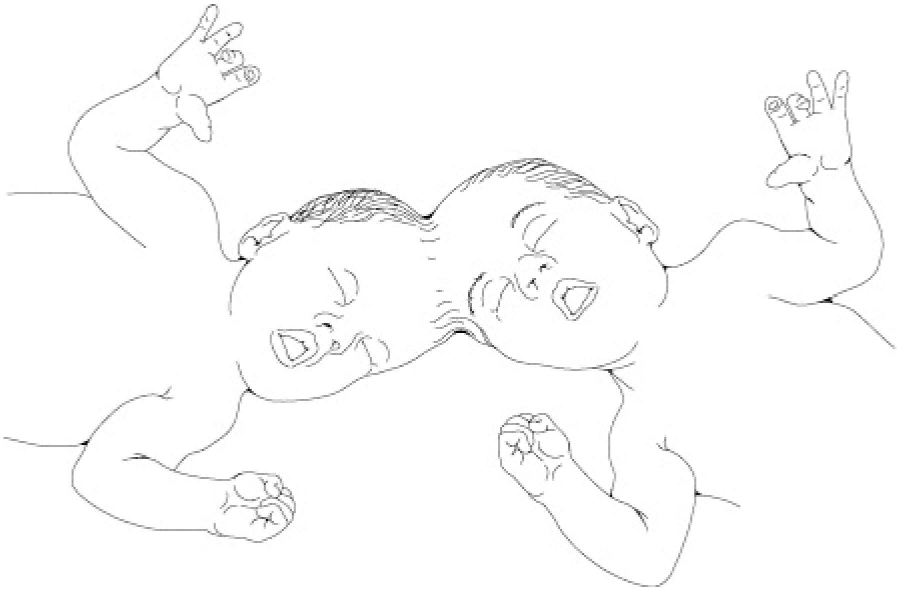
The twins may lie face to face or end to end (**Figure 92.4**). They have two sacra or two symphysis pubis. They may share the lower GI tract (70%) and/or the GU tract (50%) and may have crossing ureters. The twins can be tetrapus, tripus, or bipus, although the most common arrangement is the presence of four legs. Pelvic conjunction leads to complex urogenital and orthopedic anatomy. The kidneys usually function normally, but are often malrotated or ectopic in location. When two bladders are present, they lie side by side in a collateral position or they may lie in a sagittal midline location with one bladder draining into the other. The ureters frequently cross over and insert into a contralateral bladder, such that they will need to be rerouted during separation. Partial urethral duplication is possible, but a single urethral orifice is typical. The distal GI tract is often shared, with anorectal agenesis and rectovesical fistula. Contrast studies are necessary to delineate distal bowel anatomy. Urogenital sinus or cloaca may be present. In boys, there is an increased incidence of undescended testes.



92.4 Ischiopagus.

Craniopagus

The twins may be joined at any part of the skull and share the skull, meninges, and the venous sinuses (**Figure 92.5**). The brains are usually separate, although some cortical fusion can occur in 33% of cases.



92.5 Craniopagus.

Cephalopagus

The twins often have a fused thorax in addition to a fused head. The single fused head may have two faces (janiceps) facing away from each other; one face may be rudimentary. These twins often die *in utero* or shortly after birth as they are non-viable.

Rachipagus

Rachipagus is extremely rare. The twins are joined dorsally and face away from each other. The fusion terminates above the sacrum. The twins have generally vertebral anomalies and neural tube defects. Separation is not usually considered.

PRENATAL DIAGNOSIS

The diagnosis of conjoined twins should be considered in any twin pregnancy that has a single placenta and no visible separating amniotic membrane. Polyhydramnios occurs in as many as 50% of conjoined twin pregnancies compared with 10% of normal twins and 2% of singleton pregnancies.

Prenatal ultrasonography (US) is capable of diagnosing conjoined twin pregnancies as early as 12 weeks' gestation. The sonographic findings include inseparable fetal bodies and skin contours, an unchanged relative position of the fetuses, both fetal heads persistently at the same level, and a single umbilical cord containing more than three vessels. The addition of color Doppler and 3D US has aided earlier and more accurate diagnosis. Detailed scanning at around 20 weeks' gestation will accurately define the extent of the conjoined area and provide an assessment of which viscera are shared. Fetal echocardiography is mandatory, as twins with a complex shared heart have an extremely poor prognosis and termination of the pregnancy is invariably recommended. Computed tomography (CT) and magnetic resonance imaging (MRI) may be performed at 32–34 weeks' gestation, but once the decision has been made to proceed with the pregnancy, these investigations can be carried out with greater accuracy after delivery. An additional advantage of prenatal diagnosis is that the time, place, and mode of delivery can be planned. The delivery should take place at or close to the surgical unit where separation will be performed. Delivery must always be by Cesarean section at 36–38 weeks' gestation.

MANAGEMENT

The management of conjoined twins can be divided into four separate time frames: Prenatal, non-operative treatment, emergency separation, and elective separation.

Prenatal

Once the diagnosis of conjoined twins is suspected on prenatal scan at 12 weeks' gestation, it is important to define the anatomy of the union. Termination of the pregnancy is recommended where there is complex cardiac fusion or extensive cerebral fusion. Detailed echocardiography and accurate US is essential. The extent of deformity expected following possible subsequent separation must be carefully and accurately explained so that an informed decision can be made either to terminate or to proceed with the pregnancy.

Non-operative treatment

No attempt at surgical separation should be considered in the presence of complex cardiac or cerebral fusion or where the expected deformity following separation is unacceptable to the parents.

Emergency separation

This procedure is undertaken when one twin is dead or dying and threatening the survival of the remaining twin, or where a life-threatening correctable congenital abnormality (e.g. intestinal atresia, malrotation with or without volvulus, ruptured omphalocele, or anorectal agenesis) is present in one or both twins. In these circumstances, the only chance of saving one or both infants lies in immediate separation. Emergency separation carries a significantly higher mortality rate compared with elective procedures.

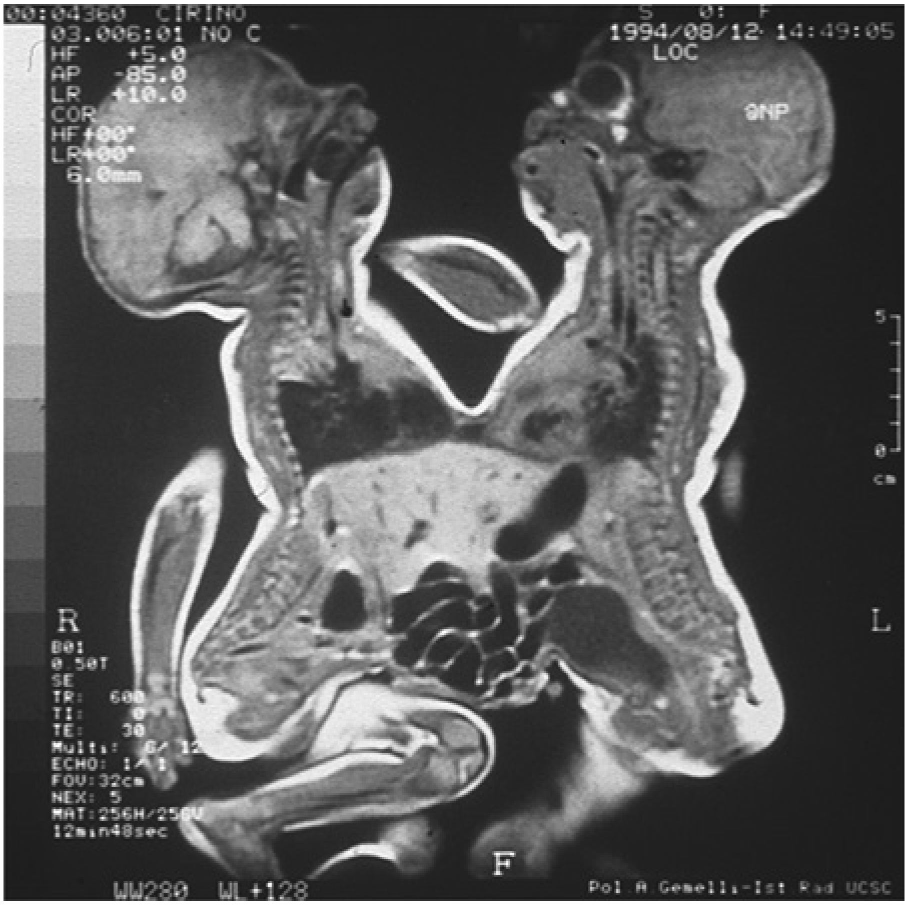
Elective separation

This will normally take place between 2 and 4 months of age. It allows the twins to stabilize and thrive, and provides time to carry out detailed investigations to define the nature and the extent of union. It also allows the application of methods to be carried out to achieve primary closure of the wound, such as tissue expansion. Detailed planning of the operative procedure with all members of the operating team, nursing staff, intensive care, laboratory staff, and imaging unit as well as administration should take place before the separation.

The survival rate for elective separation is in excess of 80%.

INVESTIGATIONS

The choice of imaging study will depend on the site of union. For thoraco-omphagopagus twins, essential investigations include echocardiogram, CT (**Figure 92.6**), and MRI scans with particular attention directed to the anatomy of the hearts, livers, and GU systems. Where the livers are fused, it is important to document the presence of separate gall bladders and hepatic veins. GI contrast studies are useful in showing separate GI systems. MRI angiography has superseded percutaneous angiography to define vascular anatomy. Bony anatomy is best demonstrated on plain X-ray and MRI scan.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92.6 CT scan of omphalo-thoracopagus twins.

ANESTHETIC MANAGEMENT

Two sets of anesthesiologists, one for each infant, are essential, as each has to be separately monitored throughout the procedure. Essential monitoring consists of arterial and central venous catheters, electrocardiogram, pulse oximetry, capnography, and urinary output. All lines need to be color-coded for individual infants to avoid confusion when their position is changed. Regular blood gas analyses are undertaken throughout the procedure. All drugs and IV fluids are calculated on a total weight basis, with half being delivered to each twin. Because of the cross-circulation, drugs given intravenously may have an unpredictable effect. Thus, particular care is essential when administering drugs such as opioids, which should be given incrementally.

OPERATIVE PROCEDURE

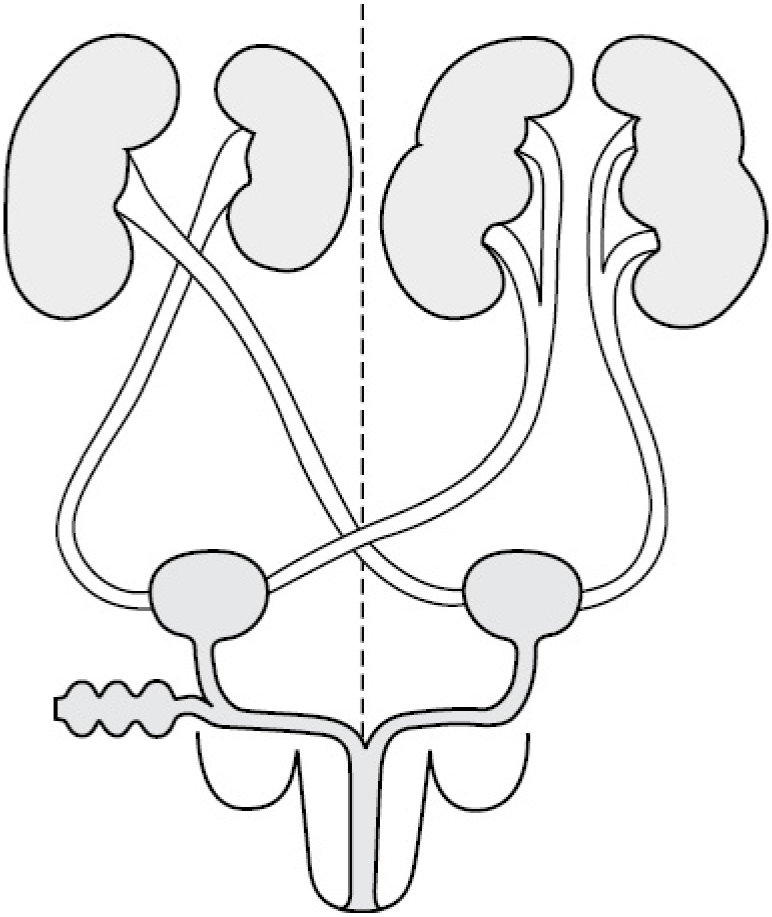
Technical details of the operative procedure will be dictated by the anatomy of the junction and by the organs and structures shared.

In thoracopagus, the liver is invariably shared. In 90% of cases, there is a common pericardium which can be separated to provide an individual pericardial sac for each twin. Major myocardial connections are present in 75% of cases and only a few attempts have been made at separation. The upper GI tract is common in 50% of cases with a shared biliary system in 25%.

In omphalopagus, the liver is shared in 80% of cases and in 33% the intestines join in the third part of the duodenum and separate at the level of the Meckel's diverticulum.

When the livers are fused, the plane of division may be obvious. In the absence of a clear plane for division, a useful maneuver is to develop a plane behind the two main masses of liver and to use this bulk of liver to digitally compress each half during separation with ultrasonic dissection and bipolar cautery to limit blood loss and seal vessels. The raw surfaces of the divided livers are sealed with fibrin sealant.

In pygopagus, ischiopagus, and parapagus, the intestines are separate to the level of the Meckel's diverticulum. The common terminal ileum and colon have a dual blood supply. The GU tract is shared in 15% of the former and in 50–100% of the latter two types. It is not uncommon for the ureters in these situations to cross over from one twin and enter the contralateral bladder (**Figure 92.7**).



92.7 Urological anatomy in one set of conjoined twins.

Blood loss may be a major intraoperative problem, especially where there is pelvic fusion.

Despite every attempt to define as accurately as possible all anatomical connections prior to surgery, "unexpected events" are frequently encountered during the operation. Examples in our experience include abnormal vascular communications, and previously unidentified intestinal and GU anomalies. The surgical team should be aware of these variations in anatomy and be prepared to vary the operative procedure accordingly.

When primary closure of the defect proves impossible despite all possible maneuvers, it will be necessary to insert prosthetic material (polypropylene mesh, Silastic sheet, Gore-Tex®) as a temporary measure. The insertion of a prosthetic patch in closure of the abdomen is preferable to "closure under tension," which may embarrass respiration or inhibit venous return.

POSTOPERATIVE CARE

Postoperatively, all surviving infants are extremely fragile.

All intraoperative monitoring must be continued postoperatively in the intensive care unit and because of the prolonged duration of surgery, the infants are electively paralyzed and mechanically ventilated for a variable period of time.

Meticulous attention should be directed at monitoring fluid and electrolyte balance, and in particular avoiding over-hydration which may precipitate cardiovascular instability. Sepsis is a major cause of mortality and morbidity and strict infectious precautions must be exercised, particularly where large skin defects are present. Progressive tucks of the prosthetic mesh should be instituted early in the postoperative period with the expectation that full closure will occur within a week or two.

Late unexpected deaths following separation are unfortunately not uncommon. One twin is fragile and is dependent on survival being joined to its twin.

OUTCOME FOR CONJOINED TWINS TREATED AT GREAT ORMOND STREET HOSPITAL

Since 1985, we have had experience with 38 pairs of conjoined twins at Great Ormond Street Hospital in London:

- *Non-operative treatment* was carried out in nine sets, all of whom died within a short period of time.
- *Emergency separation* was attempted in 11 sets, with 8 survivors (33%).

- *Planned separation* was performed in 18 sets with 33 long-term survivors (89%). Of the three deaths, two occurred many months after successful separation.

Comparison with other series in the literature is shown in **Table 92.2**.

Table 92.2 **Outcome of management of conjoined twins in the major series**

Authors (Country)	Year	Number	No operation	Emergency separation (survivors (%))	Planned separation (survivors (%))
O'Neill (United States)	1988	18	5	5 sets (1 survived (10%))	8 sets (13 survived (81%))
Cywes/Rode (South Africa)	2006	33	16	None	17 sets (22 survived (65%))
Al Rabeeah (Saudi Arabia)	2006	29	19 (1 abandoned operation)	None	10 sets (19 survived (95%))
Saguil (Philippines)	2009	21	6	6 sets (1 survived (8%))	9 sets (15 survived (83%))
Spitz/Kiely/Pierro (UK)	2010	38	9	11 sets (8 survived (33%))	18 sets (33 survived (89%))

FURTHER READING

Kingston CA, McHugh K, Kumaradevan J, et al. Imaging in the preoperative assessment of conjoined twins. *Radiographics* 2001; 21: 1187–208.

Spitz L, Kiely EM. The management of conjoined twins: The Great Ormond Street experience. *Semin Pediatr Surg* 2015; 24(5): 201–2.

Spitz L, Kiely EM. Experience in the management of conjoined twins. *Br J Surg* 2002; 89: 1188–92.

Spitz L, Kiely EM. Conjoined twins. *J Am Med Assoc* 2002; 289: 1307–10.

Wilcox DT, Quinn SM, Spitz L, et al. Urological problems in conjoined twins. *Br J Urol* 1998; 81: 905–10.

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